ALVEOLAR SOFT PART SARCOMA

A REPORT OF A CASE AND A REVIEW OF THE CASES REPORTED IN THE ENGLISH LITERATURE.

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This rare and malignant tumour is unique in its histology and histogenesis. Its histology is unique because of its most characteristic and easily recognisable morphology and having studied one tumour or seen good micro-photographs of one, a pathologist will find no difficulty in identifying another. This tumour has been intensively studied for the last 30 years, and yet we are now just as ignorant as in 1934 (Klemperer) about its histogenesis. Two other names have been constantly used for this category of tumours:- (1) Malignant granular cell myoblastoma and (2) Malignant non-chromaffin paraganglioma.

It is our intention to report the first case of this tumour occurring in a scalp of a Chinese girl and to review the available English literature on this tumour with special reference to its histogenesis.

CASE REPORT

H.W.L., female Chinese, aged 18 years was first seen on 17th May, 1960 with a history of over a year's swelling in the left fronto-parietal region. This swelling was about 2 cms. in diameter, conical in shape, non-compressible, lobulated and painless. There was no history of trauma and no other symptoms. On 7th June, 1960, the tumour was excised. There was no evidence of bony involvement. The tumour was diagnosed as an atypical myoepithelioma.

On 17th November 1962, she returned again complaining of cough and fever. A chest X-ray was done and it showed a left mediastinal tumour (Fig. 1). On closer questioning she gave a history of intermittent cough for 2 years, occasional haemoptysis for a year with some loss of weight and fever for 6 months.

At operation on 7th December 1962, a tumour was found in the mediastinum, about 11 cms. maximum diameter, extending from the hilum of the left lung into the fissure between the upper lobe and the lingula and closely adherent to the upper lobe bronchus. The tumour was peeled off from the lung but the medial portion was firmly adherent to the left upper lobe, which was consequently removed as well. The tumour was diagnosed as a metastatic alveolar soft part sarcoma, and on review of scalp tumour, that proved to be the primary.

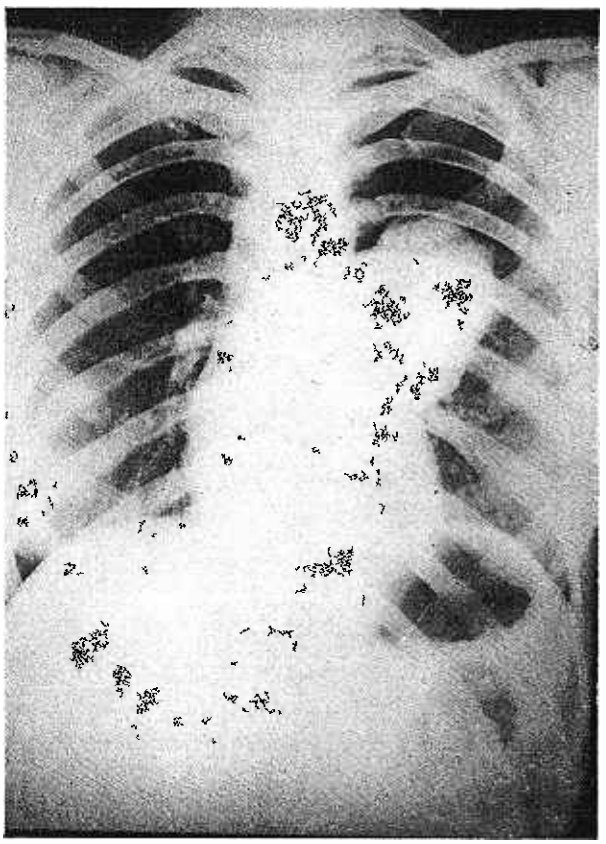

Fig. 1. Chest X-ray. The left mediastinal tumour is well shown.

A close search post-operatively revealed no other metastasis and no recurrence in the original scalp lesion.

In January 1965, multiple secondaries were found in both lungs. No treatment was being given and patient was alive at the time of writing this paper.

GROSS DESCRIPTION

The primary tumour in the scalp measured about 2 cms., in maximum diameter. It was rounded and appeared encapsulated and was situated in the dermis and subcutaneous tissue of the scalp (Fig. 4). There was no muscle in the vicinity of the tumour.

The lung secondary was again rounded and encapsulated and measured approximately 11 × 7 × 6 cms. firmly adherent to the upper lobe of the right lung. It was nodular on the outer surface (Fig. 2). The cut surface was yellowish grey with blotches of haemorrhage. Cystic spaces were also seen (Fig. 3). The consistency was firm and friable.

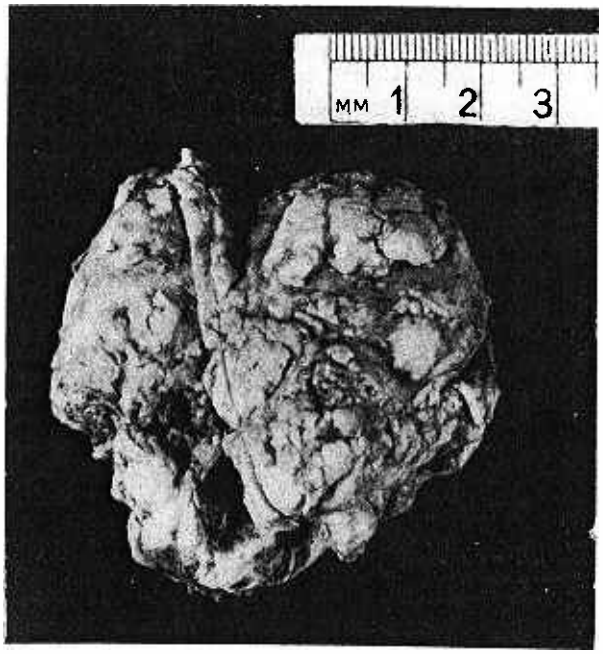

Fig. 2. The external surface shows a well encapsulated tumour with a nodular surface.

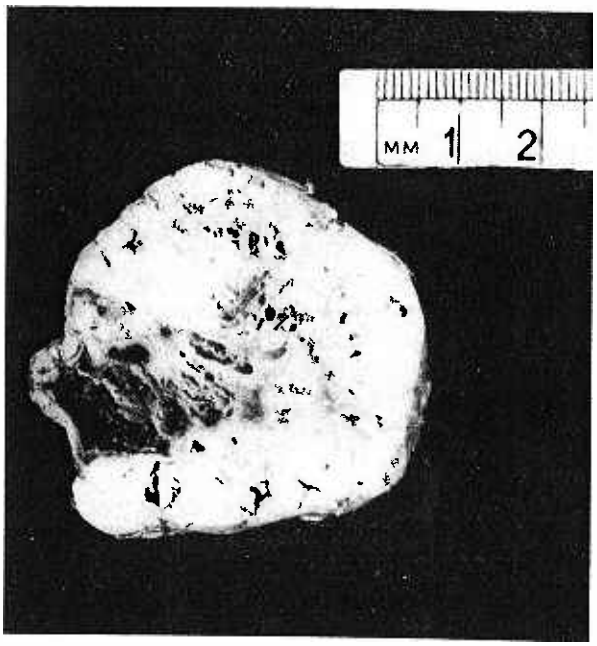

Fig. 3. The cut surface is firm, yellowish gray with areas of haemorrhage and cystic changes.

MICROSCOPIC DESCRIPTION

The tumour masses were composed of discrete solid cords of epitheloid cells completely

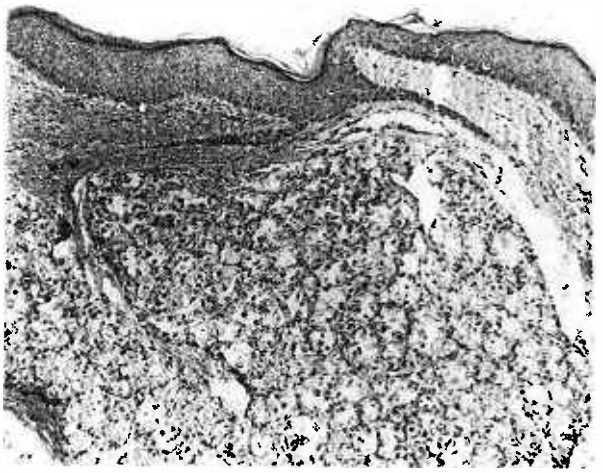

Fig. 4. Typical alveolar soft part sarcoma almost encroaching upon the epidermis.

invested by thin-walled vascular channels (Fig. 5). These cords of tumour cells were for the most part jostled tightly together except in an occasional area where a dilated vascular channel was being invaginated by the tumour cords. The vascular channels were mainly composed of a layer of endothelium backed by a thin layer of reticulin (Figs. 6 and 12). They quite often contained blood elements. The cords of tumour cells varied in number between two to 200 (Fig. 7), and except for an occasional isthmus joining one "island" to a larger "island" (Fig. 8), the cords were invariably separated from the neighbouring cords by capillaries (Fig. 9). This tumour-vascular complex was characteristic of this tumour.

The central portions of the cords became degenerated and fell away; thus a pseudo-alveolar pattern was created (Fig. 10).

The individual cells were mostly large and polyhedral in character; sometimes they were rounded or oval. They had indistinct cellular outlines (Figs. 9, 10 & 11). They vary in sizes between 10 to 50 u with an average of 25 u. The cytoplasm of the tumour cells was eosinophilic and was either finely granular or vacuolated (Figs. 9 and 11). The former type cell was more often found at the periphery of the tumour masses and the vacuolated ones were more generally found centrally (Figs. 10 and 11). The impression was that the vacuolated forms were degenerating tumour cells.

Homogenization and condensation of the cytoplasm occurred frequently and these were interpreted as "inclusion bodies" by some authors (Figs. 10 and 11).

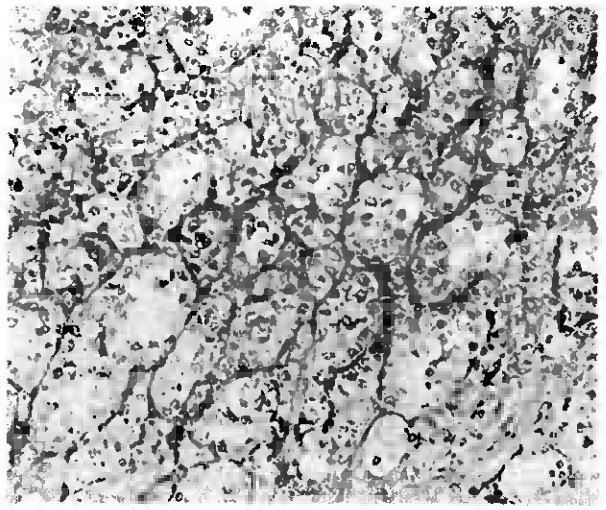

Fig. 5. Solid cords of epitheloid cells invested by capillaries (X 150).

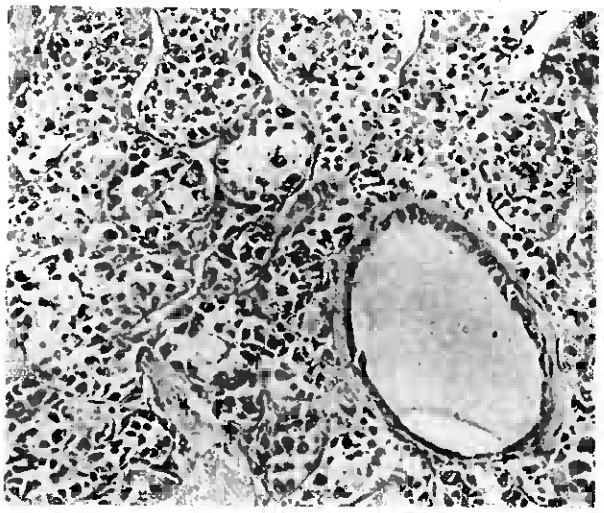

Fig. 8. The capillaries or sinusoids are more evident here. The large tubular structure is a bronchiole (X 50).

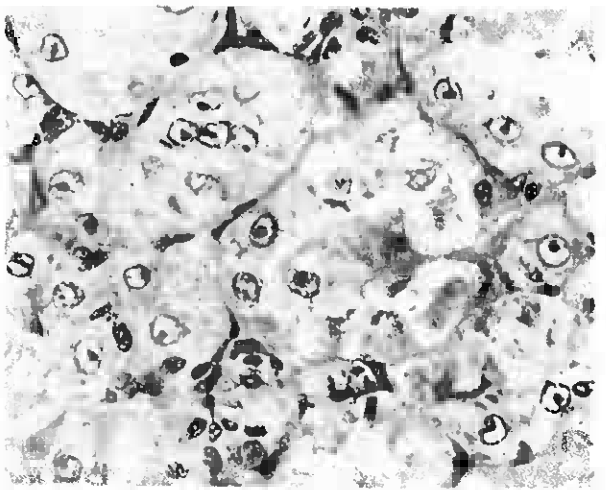

Fig. 6. High power of Fig. 5 showing the tumour-vascular relationship (X 500).

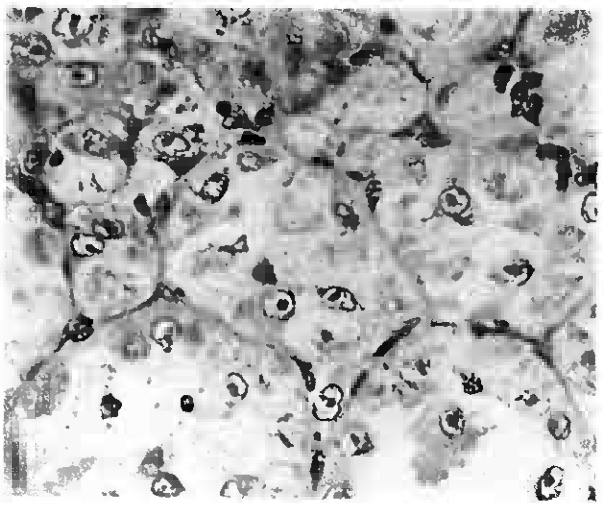

Fig. 9. The cytology shows poor cellular outline with a granular or vacuolated cytoplasm. Prominent nucleoli with relatively clear cytoplasm (X 500).

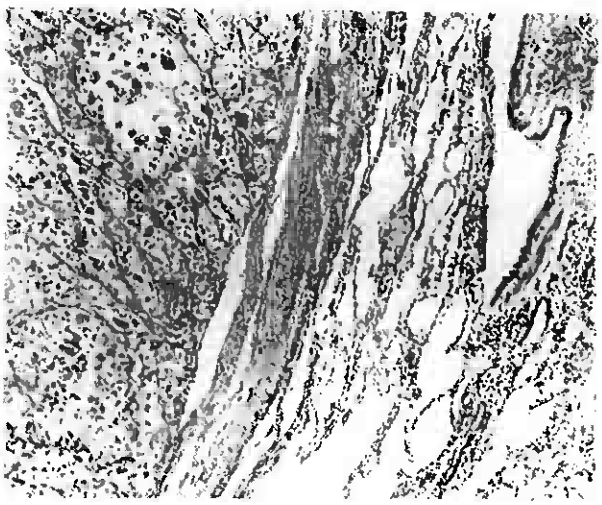

Fig. 7. Well encapsulated secondary in the lung (X 75).

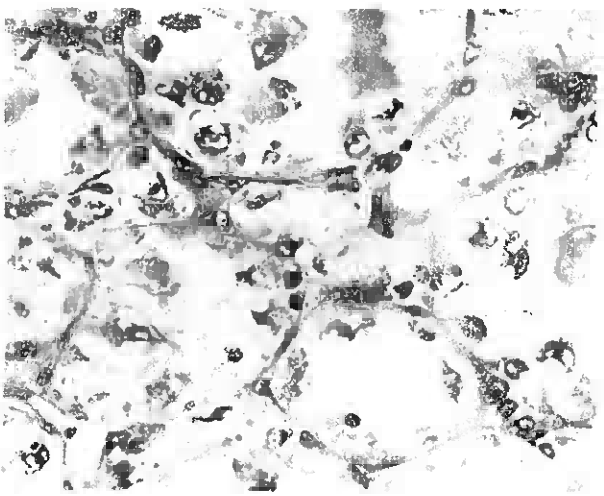

Fig. 10. Note pseudo-alveolar pattern due to fall out of degenerated cells. Homogenised cytoplasm is seen at top with no nucleus in the cell (X 500).

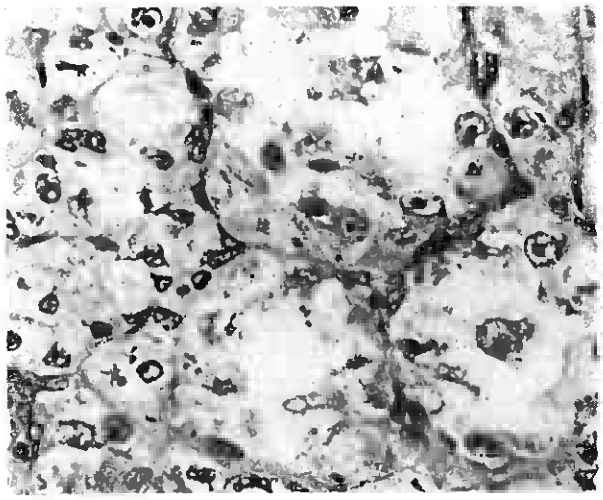

Fig. 11. Vacuolated cytoplasm is well shown here (X 500).

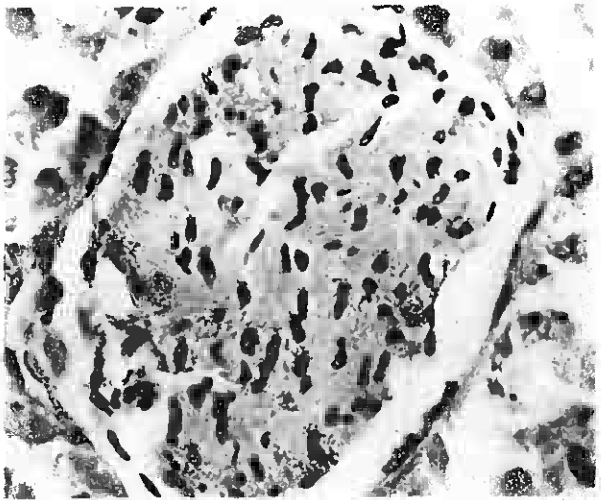

Fig. 14. Normal nerve within metastatic alveolar soft part sarcoma in lung (X 500).

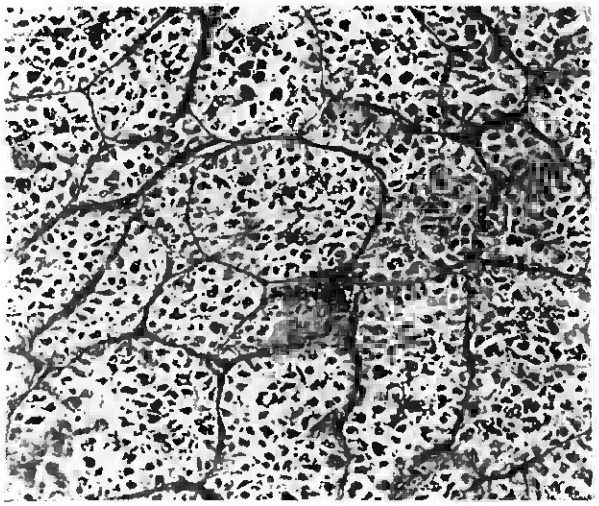

Fig. 12. Distinct reticulin surrounding cords of cells but fibres are not seen within the cords (X 150).

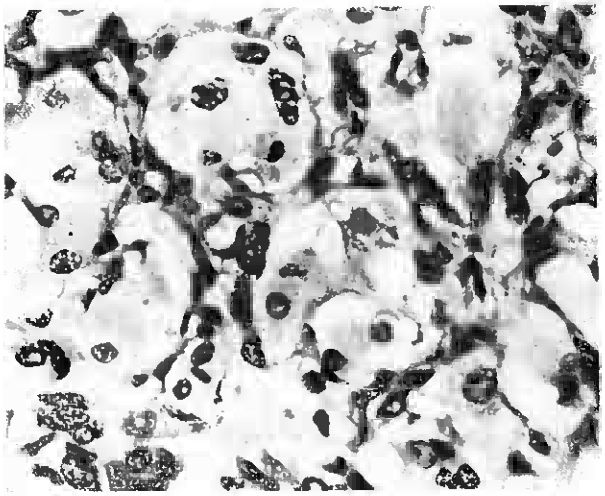

Fig. 15. Carotid body tumour. Compare with Figs. 9, 10 & 11. The smaller groups, the finer cytoplasm and the less malignant nuclei are obviously contrasts (X 500).

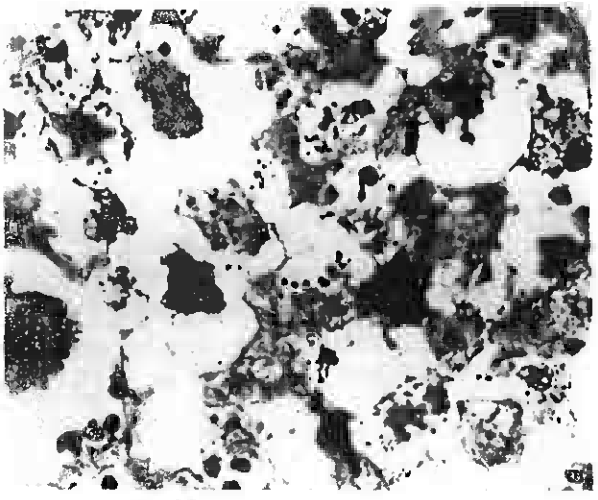

Fig. 13. These P.A.S. positive granules are diastase resistant (X 500).

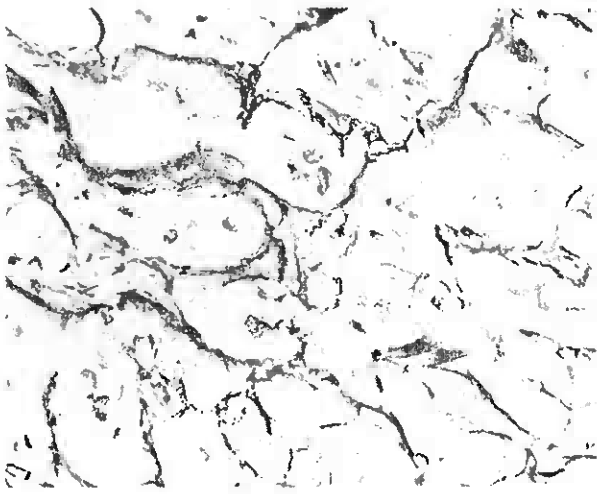

Fig. 16. The reticulin is not as definitive and not as complete in its encircling of the tumour groups (X 500).

TABLE I

LIST OF PUBLISHED CASES OF ALVEOLAR SOFT PART SARCOMA

Authors	No. of Cases	Year	Name Used
Klemperer	1	1934	G.C. Myoblastoma (Case 6 Fig. 10)
Hartz	1	1944	G.C. Myoblastoma
Ackerman & Phelps	1	1946	Malignant Myoblastoma
Khanolkar	1	1947	Malignant Myoblastoma (Case 1)
Schwidde, Meyers & Sweeney	1	1951	Metastatic G.C. Myoblastoma
Smetana & Scott	14	1951	Malignant Non-chromaffin paraganglioma
Christopherson, Foote & Stewart	12	1952	Alveolar Soft Part Sarcoma
Randall & Walter	1	1954	Malignant Non-chromaffin paraganglioma
Sirsat	1	1954	Malignant Non-chromaffin paraganglioma
Hicks & Leitch	1	1955	Malignant Non-chromaffin paraganglioma
Hurley	3	1956	Alveolar Soft Part Sarcoma
Fisher	1	1956	Alveolar Soft Part Sarcoma
MacFarlane & MacGregor	1	1958	Malignant Non-chromaffin paraganglioma
Farquharson	1	1960	Alveolar Soft Part Sarcoma
Karnauchow & Magner	3	1963	Alveolar Soft Part Sarcoma
Vakil & Sirsat	10	1963	Alveolar Soft Part Sarcoma
Tan & Choo	1	1965	Alveolar Soft Part Sarcoma
	Total = 54		

TABLE II

LIST OF CASES OF ALVEOLAR SOFT PART SARCOMAS QUOTED BY AUTHORS

Author	No. of Cases	Year	Quoted By
Horn & Stout	4	1943	Horn & Stout
Masson	2	1951	Christopherson et al
Marshall & Horn	3	1961	Marshall & Horn
Willis	5	1953	Willis
	Total = 14		

TABLE III

Author	Sex	Age	Race	Primary Site	Secondaries	Gross description of primary tumour	Trauma	Course and Follow-up
1. Klemperer	F	42	Not mentioned	Calf	Nil	7·5 cm. in diameter. Partly encapsulated. C/s fleshy, pale brown	No	Excised. Well. No recurrence or metastasis 10 months later.
2. Hartz	M	40	Negro	(R) thigh muscle	Nil	8 × 5 cm. Incompletely encapsulated. C/s grey.	No	Well 2 years after operation No good follow-up.
3. Ackerman & Phelps	F	21	Caucasian	(R) gluteal region	Lymph nodes. Lungs. Bones.	Not mentioned, but recurrence at same site was huge.	Yes	Recurrence with multiple metastases 3 years later and patient died 5 years later.
4. Khanolkar	F	32	Sinhalese	(R) gluteal muscle.	Recurrence. Anxillary region.	7 × 6 cm. Encapsulated. Firm. C/s greyish-yellow divided by bands.	Yes	Recurrence and metastasis occurred 8 months after operation. Radiotherapy. Died 10 months after operation.
5. Schwidde et al	F	21	Not mentioned (? Caucasian)	(R) thigh muscle.	(R) Parietal lobe of brain. (R) humerus.	10 × 5 × 4 cm. Encapsulated (incomplete). Surrounding muscle infiltrated.	No	7 months after operation, brain metastasis removed. 37 months later a secondary in neck of humerus was found. Radiotherapy. Alive.
6. Smetana & Scott	M	27	Caucasian	(R) thigh muscle.	Lungs, pelvic bones & retroperitoneal lymph nodes.	24 × 18 cm. Partially encapsulated. Soft, pink with fibrotic necrotic areas.	Yes (Optn)	Excision and X-ray. Died 3 years after onset with local recurrences and metastases. No response to X-ray.
7. Smetana & Scott	M	29	Caucasian	(R) thigh muscle	Lungs, parietal and frontal bones, tissue of (R) shoulder	8 × 4·5 × 3·3 cm.; well encapsulated, poorly lobulated, dirty grey.	No	Excision and X-ray. Recurred and living 96 months later with metastases. No response to X-ray.
8. Smetana & Scott	M	28	Caucasian	(R) fossa ovalis	(L) side of back above hip.	5·5 × 5 × 3·5 cm. Non-encapsulated, granular yellow-brown.	Yes (Optn)	Excision. Living with metastasis 78 months after recognition of disease.
9. Smetana & Scott	M	36	Caucasian	(L) thigh	Lung and brain	8 × 6 cm. Encapsulated, lobulated, firm and rubbery, yellow-white	Yes	Excision. Living with metastases 36 months later.
10. Smetana & Scott	F	26	Caucasian	(L) popliteal fossa	Lung	11 × 7 × 5 cm.; encapsulated, soft, yellow gray.	No	Excision. Living with metastases 48 months later.

11. Smetana & Scott	M	29	Caucasian	(R) thigh	No follow-up	10·5 × 9 cm.; encapsulated but shelled out readily, pale gray with brown green areas.	No	Excision. No follow-up.
12. Smetana & Scott	F	40	Caucasian	(R) thigh	No follow-up.	8 × 4 × 6 cm.; encapsulated firm, friable, nodular L red-tan to light yellow	Yes (Optn)	Excision. No follow-up.
13. Smetana & Scott	M	23	Negro	Retroperi-toneal	Lungs & adrenals.	20 × 12 × 6 cm.; poorly encapsulated, attached to psoas and sacrum, grayish-white with cystic foci and yellow areas.	No	Died 21 months after appearance of illness. X-ray.
14. Smetana & Scott	F	27	Caucasian	Retroperi-toneal	Lungs & liver	8 cm. in diameter, embedded in muscle, nodular, yellow and white	No	Excision. Local recurrences within 1 year and died 51 months after first appearance of tumour
15. Smetana & Scott	F	9	Negro	Retroperi-toneal	No follow-up.	8·3 × 6·5 × 5·5 cm.; encapsulated, lobulated, soft friable; centrally yellow and peripherally reddish purple with gray streaks	No	No follow-up. Excision 3 months after occurrence of tumour.
16. Smetana & Scott	F	39	Caucasian	Retroperi-toneal	No follow-up	29 × 15 × 9 cm.; encapsulated, cystic; fibrous trabeculae, gray with necrotic areas.	No	Excision 24 months after occurrence of tumour. No follow-up.
17. Smetana & Scott	M	21	Caucasian	(L) thigh	Lungs, liver, heart, pancreas ribs, vertebrae, lymph nodes	Liver metastasis meas. 10 cm. in diameter; soft, friable, necrotic, pale, pink. Primary in (L) thigh invaded ilium and metabulum.	No	Excision. Died 4 months after recognition of tumour.
18. Smetana & Scott	M	27	Caucasian	(R) neck muscle	Lungs, heart, brain.	Weighed 51 gm. Partially encapsulated, indistinctly lobulated, gray to pink with necrotic areas.	Yes	Excision 15 months first occurrence of tumour. Died 27 months after initial lump was recognised.
19. Smetana & Scott	F	28	Caucasian	Angle of (L) mandible (soft tissue)	Nil	1·5 × 1·5 × 1 cm. Well encapsulated.	Yes X-ray	3 years before had swollen gland at same site, which subsided with irradiation. No evidence of recurrence 57 months after initial symptom. Alive.
20. Christopher-son et al	F	20	Not known	(R) calf. Tibialis posterior muscle	Lung	13 × 6 × 4 cm. Attached to septum. Circumscribed and lobulated infiltrating bones.	Yes	Patient died 7 years later with lung secondaries.

TABLE III (continued)

21. Christopherson et al	F	38	Not known	(R) upper arm	Brain, lung, bone, skin. Recurrence.	No measurements. Encapsulated, attached to intermuscular septum	No	Recurrence 10 months after excision. Died 2 years later with multiple secondaries.
22. Christopherson et al	F	22	Not known	(R) lower abdominal wall	Lung	No description.	No	Recurrence involving caecum was removed 2 years later. Pulmonary metastasis 3 years later.
23. Christopherson et al	M	30	Not known	(R) thigh	Lungs, lymph nodes, brain, latissimus dorsi.	23 × 10 × 10 cm. Encapsulated except attachment to muscle.	Yes	Lump appeared 15 years ago after a blow. 14 years after appearance of thigh tumour, pulmonary metastases detected.
24. Christopherson et al	M	30	Not known	(R) thigh	Lung.	Size of walnut	No	Lump excised 5 years after appearance. Recurred next year. Year later pulmonary metastases appeared. Died 10 years after initial appearance of lump.
25. Christopherson et al	F	17	Not known	Arm	Nil	No description	No	Well with no recurrence 5 years later.
26. Christopherson et al	F	19	Not known	(R) buttock	Lungs & liver	Size of golf ball	No	Secondaries detected 18 years later. Alive.
27. Christopherson et al	F	12	Not known	Base of tongue	Nil	5 cm. in diameter.	No	Well with no recurrence 5 years later.
28. Christopherson et al	F	1½	Not known	Beneath triceps	Nil	4 cm. in diameter. Poorly encapsulated. C/s uniform, friable and buffy-gray.	No	Well with no recurrence 10 years later.
29. Christopherson et al	F	22	Not known	(L) thigh (rectus-femoris)	Nil	8 × 5·5 × 3 cm. Encapsulated and nodular. C/s firm and yellow-gray with bands.	No	Well with no recurrence 12 years later.
30. Christopherson et al	F	18	Not known	Forearm muscle	Nil	7 × 4·5 × 3·5 cm. Nodular and encapsulated. C/s dense and yellow.	No	Well with no recurrence 18 months later.
31. Christopherson et al	F	19	Not known	(R) thigh	Nil	3 cm. in diameter. Firm and rubbery.	Yes	Well with no recurrence.
32. Randall & Walter	F	15	Not known	(R) thigh (vasti muscle)	Lungs	10·5 × 5 × 2 cm. Circumscribed.	No	One-third of tumour was removed 7 years after appearance. Radiotherapy was of no use. Completely excised 2 months later. Secondaries in lung removed 3 months after symptom, alive with secondaries 9 years later.

33. Sirsat	F	38	Indian	(R) foot	Lung	Globular mass invading 3rd, 4th & 5th metatarsals.	No	Died 3 years later.
34. Hicks & Leitch	M	35	Caucasian	(L) thigh muscle	Nil	7 × 6 × 5 cm. Encapsulated but attached to muscle. C/s yellow with haemorrhage.	Yes	Radiotherapy. Well with no follow-up stated. Excision performed 7 years after appearance of lump.
35. Hurley	F	15	Not stated	(L) thigh muscle	Nil	7·5 × 7·5 × 5 cm. Encapsulated. C/s white	No	Lump present for 6 years before excision. Well 5 years after operation. Total 11 years.
36. Hurley	M	32	Not stated	(L) forearm muscle	Recurred. Lungs Bones	2 tumours: 7·5 × 6 cm. and 2 × 1·5 cm.	Yes	Kicked by sheep. Then haematoma drained and lump appeared. Secondaries in lung 1 year later. 3 years after operation, secondary in bones. Died almost 4 years after injury. Radiotherapy.
37. Hurley	F	9	Not stated	(L) triceps	Nil	6 × 4 cm.	No	Radiotherapy. Well with no recurrence 7 years later.
38. Fisher	M	19	Caucasian	(R) thigh	Bones, lymph nodes, liver, lungs. Recurrent.	15 × 9 × 8 cm. Encapsulated Attached to muscle. C/s yellow-tan.	No	Died after 7 years of the disease.
39. MacFarlane & Mac-Gregor	M	6	Not stated	(L) thigh	Lymph nodes, liver, lungs, vertebrae, spinal cord, brain, skull, palate and adrenal.	7 × 3 × 3 cm. Circumscribed with infiltration at one point.	No	Died 1 year after appearance of tumour.
40. Farquharson	F	20	Not stated	(L) erector spinal muscle (lower back)	Lung	5 × 7·5 cm.	No	Alive 5 years later with lung secondaries.
41. Karnauchow & Magner	F	20	Not stated	(R) thigh	Nil	4·5 × 3·0 cm. Firm. Poorly circumscribed. C/s pale-gray with yellow spots.	No	Well 9 years later with no recurrence or metastases.
42. Karnauchow & Magner	F	15	Not stated	Trapezins muscle	Nil	12 × 8 cm. Firm. C/s yellowish-white, friable.	No	Well with no recurrence 5 years after appearance of tumour.

TABLE III (continued)

43. Karnauchow & Magner	F	17	Not stated	(L) forearm	Brain, muscle, heart, lungs lymph nodes, (L) Kidney, (L) adrenal, (L) ovary.	4 × 2·5 cm. C/s yellowish and pinkish brown.	No	13 years after operation secondaries at (L) deltoid muscle and lungs. The patient died a year after this (*i.e.* 14 years after initial symptom).
44. Tan & Choo	F	17	Chinese	(L) fronto-parietal scalp.	Lungs	2 cm. in diameter. Encapsulated. Lobulated C/s firm. Grayish-yellow with	No	Alive 5 years after initial symptoms with multiple secondaries in lung.
45. Vakil & Sirsat	F	24	? Indian	(L) gluteal region	(L) iliac bone	Not given	Not given	2 years later, swelling increased 3 times the original size with no treatment. X-ray shows destruction of the (L) iliac bone due to local extension. No follow-up.
46. Vakil & Sirsat	M	24	? Indian	(L) thigh	Generalised including lungs and brain	Not given.	Not given	Excision of tumour. Died 5 years later with generalised metastases.
47. Vakil & Sirsat	M	45	? Indian	(R) orbit	Nil	Not given.	Not given	Excision of tumour. Recurred after a few months. Died.
48. Vakil & Sirsat	M	24	? Indian	(R) leg	Lungs, Tibia.	Not given.	Yes	Deep X-ray therapy. Developed metastases to lungs and died.
49. Vakil & Sirsat	F	26	? Indian	(L) gluteal region	(L) latissimus dorsi, lungs, skull, bones and brain.	Not given.	Not given	Excision with radiotherapy. Developed metastases 1 year later. Died 5 years after onset of symptoms.
50. Vakil & Sirsat	M	18	? Indian	Retroperi-toneal	(L) kidney	Not given.	Not given	Excision. Recurrence. Nephrectomy performed with radiotherapy prestoperatively. Died 7 months after onset of symptoms.
51. Vakil & Sirsat	M	31	? Indian	(L) thigh	No follow up	Not given.	Not given	Excision after 4 years of symptom. No follow-up.
52. Vakil & Sirsat	M	59	? Indian	Retroperi-toneal	(R) iliac bone and lung, (R) leg.	Not given.	Not given	Irradiation. No response. Died 3 months after the onset of symptoms with metastases.
53. Vakil & Sirsat	F	10	? Indian	Tongue	Nil	Not given.	Not given	Excision. Recurred 6 months later. No follow-up.
54. Vakil & Sirsat	M	76	? Indian	Thigh	Lung	Not given.	Not given	Lungs metastases present when first seen. No follow-up.

The nuclei were on the whole eccentric and possessed clear nucleoplasm, well-formed radicular, chromatinic strands radiate from a prominent pink nucleolus (Figs. 9 and 11). Binucleated forms were sometimes seen and mitotic figures were rare.

Only in a very rare area that pleomorphic forms with multinucleated giant cells and dense hyperchromatic nuclei were seen.

The reticulin stain surrounded each tumour group, and did not send fibres within the groups of tumour cells (Fig. 12). This was again another distinct histological characteristic.

The PAS stain showed an indiscriminate but fine shower of positive granules (Fig. 13). After diastase digestion, the granules were less, but nevertheless present. Some of the condensed and homogenised "inclusion bodies" stained positive with PAS and few of these resisted the diastase treatment. They also stained brown with Masson Trichrome.

A tiny normal nerve bundle was found in the metastasis in the lung (Fig. 14). This looked most certainly a pre-existing normal nerve bundle rather than that generated by the tumour cells.

DISCUSSION

Historical Review

Hitherto there were only about 53 cases published in the English literature (Table I). However, there were a further 14 cases (Table II) on record which were merely quoted as bona fide examples of this particular tumour, but clinical and pathological details were not given. Thus, there is a total of 67 traceable cases in the English literature.

The very first case to be published was that of Klemperer's (Case 6). Although he included this case as a myoblastoma in his paper he was never satisfied with it, and on review of the sections Christopherson et al were convinced that the tumour was definitely an alveolar soft part sarcoma. Hartz reported another case as a granular cell myoblastoma of the thigh with an organoid pattern. Solitary cases were then reported by Ackerman and Phelps, Khanolkar and Schwidde, Meyers and Sweeney as malignant granular cell myoblastoma.

Seventeen years after the first case appeared in print in Klemperer's paper, Smetana and Scott reported a series of 14 cases and gave it a new name:- "Malignant Non-Chromaffin Paraganglioma". The very next year, Christopherson, Foote and Stewart described another 12 cases under a new name of Alveolar Soft Part Sarcoma. Since then these two new names have been used with almost equal frequency. Randall and Walter, Sirsat, Hicks and Leitch and MacFarlane and Macgregor reported isolated cases as malignant non-chromaffin paragangliomas. On the other hand, under the name of alveolar soft part sarcoma, Hurley and Karnauchow described 3 cases each. Fisher performed an extensive range of histochemical tests on one tumour, and Farquharson reported one more case. Both these authors used the term alveolar soft part sarcoma. Finally, Vakil and Sirsat reviewed 11 cases, one of which was reported by Sirsat as a paraganglioma in 1954. This time they concluded quite emphatically that this tumour was not related to the paraganglioma and was a distinct entity of its own. At this juncture the label alveolar soft part sarcoma was used instead.

In the space of 31 years, these 53 tumours were published under 3 names: 5 were called granular cell myoblastoma, 18 were labelled as malignant non-chromaffin paraganglioma and the remaining 30 went under the name of alveolar soft part sarcoma.

Clinical Features (See Table III)

Only 54 (including the present case) of the reported cases will be discussed (Table I).

Sex: There was a slight preponderance of females over males: out of the total 54 cases, 32 were females.

Age (Table IV): The ages vary between 1½ - 76 years. However, the majority of them (44 out of 54) occurred in the second, third and fourth decades. The average age was 25 years old.

TABLE IV

AGE DISTRIBUTION OF ALVEOLAR SOFT PART SARCOMA

Year	0-9	10-19	20-29	30-39	40-49	50-59	60-69	70-79
No.	4	13	21	10	4	1	0	1

Almost half occurred in the first 3 decades, and 88% in the first 4 decades. Thus it could be said that this tumour affected mainly adolescents and young adults.

Site (Table V): The commonest site was in the thigh, numbering 23 in all. Out of the total 54, thirty-two occurred in the lower limbs, seven in the upper limb, six in the retroperitoneal space, four in the head, two in the neck, two in the back and one in the abdominal wall.

TABLE V

LOCATIONS OF ALVEOLAR SOFT PART SARCOMA

Site	Thigh	Leg	Gluteal region	Arm	Retroperitoneal region	Head	Neck	Back	Abdominal
No. of Cases	23	4	5	7	6	4 { 2 Tongue, 1 Orbit, 1 Scalp }	2	2	1

Most of these tumours occured either within voluntary muscle or along muscle planes. The few exceptions are those in the retroperitoneal connective tissue (case 8 of Smetana and Scott and both retroperitoneal tumours of Vakil and Sirsat), one in the soft tissue beneath the angle of the left mandible (case 14 of Smetana and Scott), one in the soft tissue or the orbit (case 4 of Vakil and Sirsat) and lastly our case in the scalp.

Race: In quite a number of the papers reviewed, the racial origin was not mentioned. However, gauging from the countries where these reports came from, no race was immune from this neoplasm. All the eleven cases of Vakil and Sirsat could be assumed to be of Indian origin. Besides these, the non-Caucasian races were as follows: one Sinhalese, three Negroes and one Chinese. Assuming that all the rest were Caucasians, we have 38 Caucasians out of the 54 cases. As numerous tumours would have certainly been misdiagnosed all over the world, this slight preponderance of Caucasians must be accepted with extreme caution.

Natural History (Table VI): Eleven cases had no good follow-up. Out of the remaining 43 cases, 21 died of the disease, 13 were alive with metastases, nine were apparently well with no recurrence or metastasis at least 5 years after the discovery of the tumour. All in all the prognosis was quite poor; having an apparent cure rate of 21%.

TABLE VI

PROGNOSIS OF ALVEOLAR SOFT PART SARCOMA

No. of Cases with poor follow-up	11
No. of Cases died	21
No. of Cases alive with metastases	13
No. of Cases with no metastases (cured)	9
Total =	54

TABLE VII

DISTRIBUTION OF METASTASES OF ALVEOLAR SOFT PART SARCOMA

Site	Lungs	Bone	Central Nervous System	Muscle & Soft Tissue	Lymph nodes	Liver	Heart	Adrenal	Kidney	Ovary	Pancreas	Palate
No. of Cases	28	13	9	8	7	5	3	3	2	1	1	1

Metastases (Table VII): The most frequent site of metastasis was in the lung, occurring 28 times. Bone was a poor second, being incriminated 13 times. Next came the central nervous system which was the site of secondaries in 9 cases, with muscle and soft tissues in 8. The other sites mentioned were: lymph nodes (7), liver (5), heart (3), adrenal (3), kidney (2), pancreas (1) and palate (1).

Treatment: The only effective treatment was wide surgical excision. Irradiation was tried in a number of cases and found to be quite ineffective.

HISTOGENESIS

As suggested earlier, the histogenesis of this malignant tumour was still undetermined after 30 years of study. The possibility that it was a secondary from a primary carcinoma of the kidney, liver or an endocrine organ was readily dismissed by not finding the primary at post-mortem, and histologically the alveolar soft part sarcoma was quite distinct from any known carcinoma. Similarly, it did not resemble any known malignant tumour or fat, muscle or nerves.

The resemblance to the granular cell myoblastoma was based mainly on the eosinophilic granular cytoplasm and on the previous misconception that granular cell myoblastoma was of muscle origin. The different sites of origin and the different basic architecture of these two tumours were enough to dissociate the two completely. The alveolar soft part sarcoma had the characteristic pattern of a solid cord of tumour cells surrounded by capillary channels; this was not seen in the granular cell myoblastoma, and one certainly did not expect the malignant variety to be more orderly. The microphotographs of the three cases of so-called malignant granular cell myoblastomas reported by Ross, Miller and Foote were clearly not that of alveolar soft part sarcoma.

The most popular theory on histogenesis came from Smetana and Scott, who suggested that this tumour was a malignant non-chromaffin paraganglioma. They quoted Lent C. Johnson of having found a doubtful paraganglionic structure around the femoral vessels. Hurley quoted a further personal communication with Johnson who said that he was successful only on this one occasion in finding such structures. Karnauchow, however, had failed to detect any of these structures in the lower extremities after a systemic search in adults, children, infants and foetuses. Thus this single discovery of Johnson must be treated with the greatest suspicion. The absence or presence of these paraganglionic glomera in the lower extremity was of prime importance since 32 out of 54 occur in this region.

Another objection to the paraganglionic theory was the natural history of alveolar soft part sarcoma. Most non-chromaffin paragangliomas were benign, and Romanski could only trace 9 cases of malignant paragangliomas with distant metastases from the literature. Similarly, he also could cite only 8 cases with metastases to the regional lymph nodes. He also concluded that histologically there was no difference between the benign and malignant varieties of this tumour. On the other hand, alveolar soft part sarcoma was a highly malignant tumour and if left untreated would inexorably kill the patient.

A final discrepancy between alveolar soft part sarcoma and paraganglioma was the histology and histochemistry. Admittedly, there was a resemblance in the cytology and the tumour-vascular relationship was seen in both (Fig. 15). On the other hand, the differences greatly outweigh these 2 superficial similarities. The cords in the paraganglioma varied very little in their sizes, and the vascular investment was not as complete as in the alveolar soft part sarcoma (Figs. 15, 16). The paraganglionic tumour cell was much smaller (average 18 mu) than the cell of the alveolar soft part sarcoma (average 25 mu). The former varied between 8-36 mu (Marshall and Horn 1961) and the latter 10-50 mu. The eosinophilic granules in the paraganglioma were much finer whilst those in the alveolar soft part sarcoma were rather coarse. Degenerative changes in the form of vacuolization and eosinophilic condensates (crystalloid bodies), constant features in the alveolar soft part sarcoma, were not seen in the paraganglioma.

The most serious and valid difference between the two tumours came from histochemical studies. The alveolar soft part sarcoma gave a strongly positive reaction with PAS which was diastase resistant. There was no such reaction with the paraganglioma. Hamperl and Lattes with the modified Bodian Protargol stain showed positive argyrophilic granules in the normal carotid body and paraganglioma, but these granules were not found in the alveolar soft part sarcoma. This basic histochemical difference was of paramount importance. The other striking difference was that paraganglioma gave

positive reaction for phospholipid stains, whilst the alveolar soft part sarcoma was constantly negative (Marshall and Horn, Fisher).

In conclusion, the differences in these two tumours were too numerous to associate them histogenetically. On the other hand, no evidence could be brought forward towards an alternative histogenesis.

SUMMARY

An alveolar soft part sarcoma occurring in the scalp of a Chinese girl is reported. She is living with pulmonary metastases, 6 years after occurrence of the tumour.

The entire English literature is reviewed, and 53 other cases are found reported, whilst 14 other tumours are alluded to have been seen by various authors. The clinical aspect and natural history of the 54 reported cases are summarised.

Histogenetically, the origin of this tumour is still unsettled. The clinical, histological and histochemical differences between the paraganglioma and alveolar soft part sarcoma are too numerous for the assumption that the latter is the malignant counterpart of the former. Well documented reports of malignant paragangliomas (Romanski) show that the malignant and the benign paragangliomas do not differ histologically. In conclusion, the histogenesis of the alveolar soft part sarcoma remains unsolved.

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