A CASE OF EMBRYONAL RHABDOMYOSARCOMA WITH UNUSUAL PRESENTATION

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INTRODUCTION

Embryonal rhabdomyosarcoma is an uncommon tumour usually seen in the head and neck of young children and adolescents. It is also known to occur in sites where there is little or no striated muscle. Publications from the Memorial Center for Cancer and Allied Diseases, New York, by Stobbe and Dargeon (1950) and Moore and Grossi (1959) have recorded 52 cases during the period 1935 to 1959, in which the neoplasm was found in the head and neck of children between 7 weeks and 16 years old. Few large series in adults have been published. Lawrence *et al* (1964), however, have reviewed 48 cases of embryonal rhabdomyosarcoma seen over a 24-year period (1935-1959). In their cases they excluded those tumours that are of urogenital or head and neck origin. Although 42 of them were children from 16 days to 14 years (mean age of 4.9 years), 6 were adults between 26 and 51 years (mean age 35.3 years). Besides the head and neck region, the neoplasm is less commonly seen in the extremities, abdominal and pelvic retroperitoneum, buttocks, perineum and groin, abdominal and pelvic retroperitoneum, urogenital tract including spermatic cord and tunica vaginalis of testes, common bile duct, and mediastinum. In bone the neoplasm has been recorded to occur in the mandible (3 cases) and distal phalanx of the thumb.

In this paper we are reporting a case of embryonal rhabdomyosarcoma located in the femur of a young adult who also presented with haematological features.

CASE REPORT

Clinical History: On 14-9-67, CLS, a 21 year old Chinese male was brought to see a general practitioner with the complaints of generalised weakness of two weeks duration, and pain in the left hip. 10 days previously, while brushing his teeth he felt dizzy and fell to the floor following which he could not raise himself. The general practitioner examined him and referred him to Outram Road General Hospital as a case of fracture of the left femur and severe anaemia for investigation.

Physical Examination: His general condition was poor. He was slightly dyspnoeic and showed marked pallor, bleeding from the gums and petechial haemorrhages and ecchymoses in the skin of the lower limbs. Both liver and spleen were enlarged. Examination of the left hip showed that he had a fracture of the neck of the left femur. No other abnormality was found.

Laboratory Investigations: Hb. 3.1 gm.%; erythrocyte count 1.7 M/cmm.; total white count 18,800/cmm.; differential count—neutrophils 27%, lymphocytes 20%, metamyelocytes 29%, neutrophil myelocytes 16%, monocytes 6%, eosinophils 2%, and an occasional promyelocyte. Platelet count 2,500/cmm. Reticulocyte count 7%. Haematocrit 8%. MCV 47 cmm. MCH 18 uug. MCHC 39%.

Peripheral blood film showed the presence of a normochromic normocytic anaemia with slight anisocytosis and poikilocytosis. There was a leuco-erythroblastic reaction. Large numbers of immature white cells were present and there were about 70 nucleated red cells per 100 white cells.

Marrow aspiration from the sternum obtained little marrow tissue which was very much diluted with blood. The smears showed numerous large round malignant cells with stippled nuclei and no definite nucleoli. The cytoplasm was basophilic and vacuolated. Many mitotic figures were seen. There was little evidence of erythropoiesis. Leucopoiesis was also depressed and there was no evidence of leukemia. Megakaryocytes were absent. Reticulum and plasma cells were reduced in numbers.

Prothrombin time: 17 sec., (control 16 sec.) Bleeding time was longer than 18 min. Clotting time: 2½ min. ESR 21 mm./hr.

Serum alkaline phophatase 3.6 units. Serum acid phosphatase 2.0 units. Blood urea 43 mg.%.

Urine analysis showed no abnormality.

RADIOLOGICAL REPORT

Skull: The cranium is of normal appearance. The sella turcica is normal in size. There is no evidence of destruction of the temporal, orbital or facial bones.

Chest: No abnormal lesion detected.

Pelvis and Upper Femora: There is much destruction of the upper end of the left femur, mostly of a patchy osteolytic character. There is a pathological fracture of the left femoral neck.

PROGRESS

The patient was given transfusions of fresh blood and platelet suspensions for anaemia and thrombocytopenia. He was also started on prednisolone 20 mg. t.i.d., tetracycline, and other supportive measures. The left femur was placed in Russel's traction. The patient's condition deteriorated rapidly. He suddenly went into coma on 27-9-67 and died a few hours later.

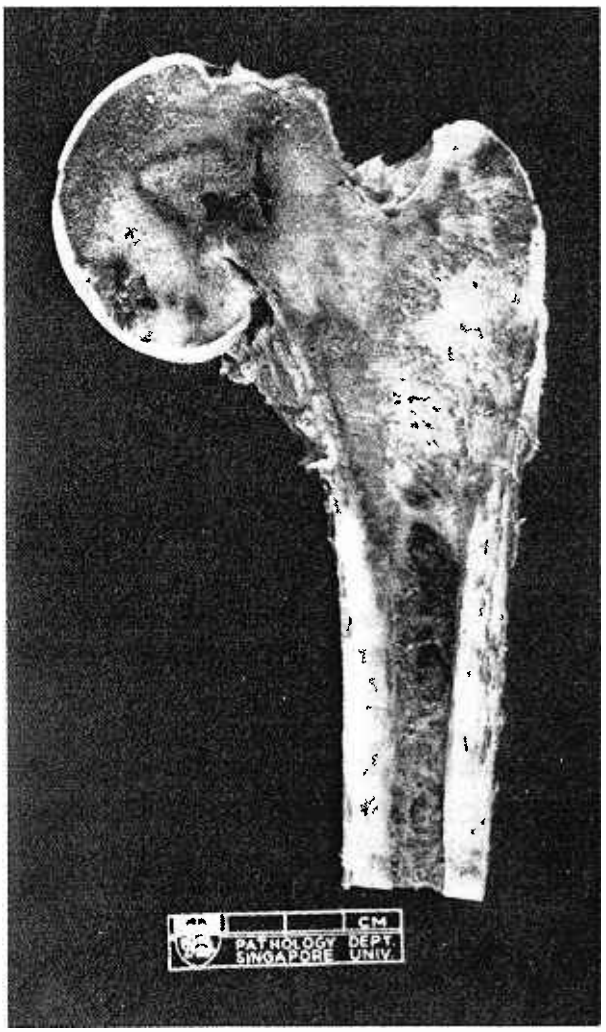

Fig. 1. Hemisection of the left femur showing location of the embryonal rhabdomyosarcoma in the upper portion of the bone. A subcapital fracture of the neck of femur is noted.

NECROPSY FINDINGS

The body was very pale and thin, and there were small areas of skin haemorrhages in all four limbs and in the trunk. No abnormal mass was found in the head and neck region or in the external body musculature. The left femur showed the presence of an irregular, greyish-white, soft gelatinous tumour (5 × 4 × 4 cm.), and there were areas of haemorrhage and necrosis (Fig. 1). The tumour had extended to the neck and head of the femur. A subcapital fracture of the femoral neck was noted. No evidence of tumour in the surrounding tissues directly outside the bone was found. Cut surface of the sternum showed small infiltrations of tumour tissue in the marrow. The heart (284 gm.) showed numerous epicardial haemorrhages and an area of subendocardial haemorrhage in the septal wall. The lungs were both congested and oedematous. The liver (1956 gm.) was slightly enlarged and pale. An area of haemorrhage was seen in the body of the pancreas. The spleen (460 gm.) was enlarged and congested. Lymph nodes were generally not enlarged. In the brain there was a large area of haemorrhage in the region of the left lentiform nucleus, internal capsule, caudate nucleus and the wall of the lateral ventricle, and extending to the left half of the midbrain. No significant findings were found in the remaining organs.

HISTOLOGICAL EXAMINATION

Sections from the tumour in the left femur show histological features of an embryonal rhabdomyosarcoma. Most of the tumour cells are round and undifferentiated, and are slightly larger than a lymphocyte. They have hyperchromatic nuclei and scanty vacuolated cytoplasm. They are seen in groups or clusters surrounded by bands of collagenous fibrous tissue or scattered about indiscriminately (Fig. 2). Oval-shaped cells, strap-shaped cells and spindle-shaped cells of varying lengths are present, the latter two types show abundant, distinct eosinophilic cytoplasm. In certain fields the spindle cells predominate and they are arranged in parallel or interlacing bands. Giant cells are few except in one field (Fig. 3), and cells with definite cross-striations are found only after careful search in sections stained with P.T.A.H. (Fig. 4).

Examination of sections from the sternum, heart, liver, pancreas, lungs, kidneys, spleen, and brain show varying degree of infiltration with the round undifferentiated malignant cells. These cells are similar to those seen in the sternal mar-

row smears. Histological examination of sections taken from muscles attached to the femur revealed no evidence of neoplasm.

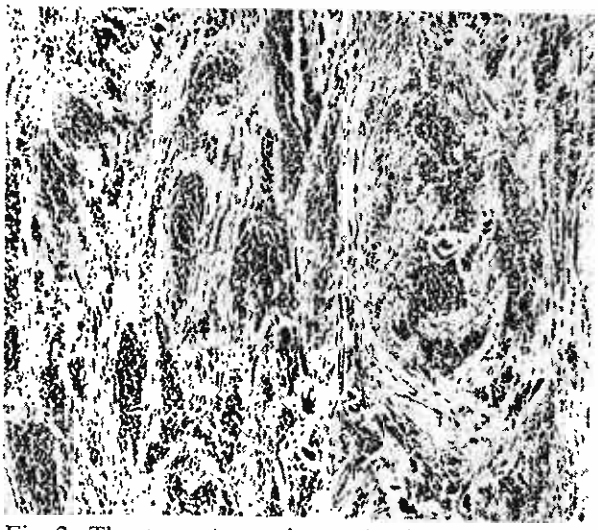

Fig. 2. This field shows the predominance of round cells arranged in clusters and surrounded by bands of collagenous fibrous tissue.

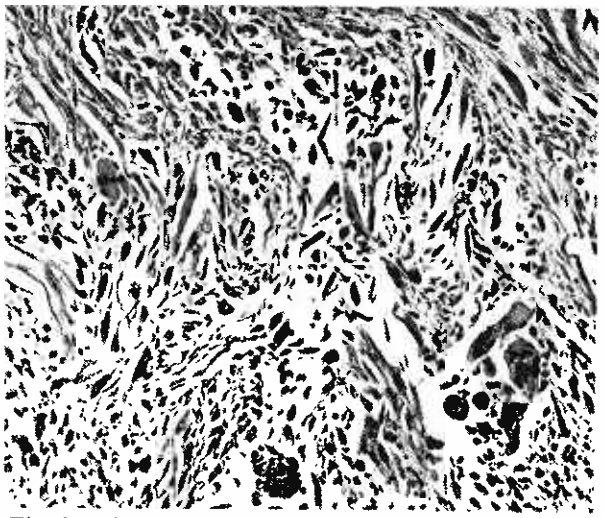

Fig. 3. Field showing spindle cells amongst oval-shaped and round cells. They are arranged in parallel or interlacing pattern. They have a single centrally placed nucleus and intensely eosinophilic cytoplasm.

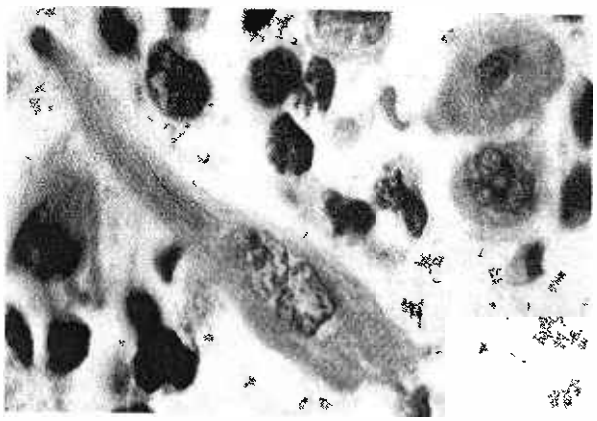

Fig. 4. "Tennis-racquet" cell with double nuclei, longitudinal myofibrils and cross-striations. Note also a large round cell at one-o'clock with vacuolated cytoplasm. The smaller round cells with hyperchromatic nuclei are completely undifferentiated and most numerous.

DISCUSSION

Since Stout's classic paper on Rhabdomyosarcoma of Muscles in 1946, STOUT (1946) pathologists began to diagnose the tumour with confidence even in the absence of the characteristic cell with cross-striations. Subsequent authours, STOBBE and DARGOON (1950), AREAN and MARCIAL-ROJAS (1957), HORN and ENTERLINE (1958), however, have found clinico-pathological differences between the adult type of rhabdomyosarcoma in Stout's cases which occur mainly in the fourth and seventh decades of life, in the skeletal muscles of the extremities, and the juvenile type which is usually seen in young children and adolescents in the head and neck region, and in structures where little or no skeletal muscle is present, such as, the middle ear, maxillary sinus, mandible, and terminal phalanx. This latter type has been called embyonal rhabdomyosarcoma. Histologically, it consists predominantly of the more immature rhabdomyoblastic cells, and cells with cross-striations and giant cells are seldom seen. It is a highly malignant tumour, rapidly fatal, and has a much poorer prognosis than its adult counterpart.

On reviewing the literature, only four cases were recorded in which the tumour had arisen in bone—three in the mandible MOORE and GROSSI (1959), and one in the terminal phalanx of the thumb LAWRENCE *et al* (1964). Its histogenesis in such sites is difficult to explain. It is believed that in these cases the tumour had arisen from remnants of undifferentiated mesodermal tissue or its derivatives.

In our case, apart from the diffuse infiltration of poorly differentiated malignant cells in the various organs, the only neoplastic mass was located in the upper end of the left femur. Radiological films of the hips and upper femora and the head and neck region revealed no lesion which could suggest a primary tumour in the skeletal muscles of these areas, or in the middle ear, nasopharynx, sinuses or orbit. Further, examination of the histological sections of muscles attached or adjacent to the upper end of the left femur failed to show neoplastic tissue. The patient moreover, did not present with the complaint of a swelling or mass, which is the usual mode of presentation in the majority of cases, even in children.

On the other hand, the probability that the femoral tumour is a metastasis cannot be completely excluded because (i) embryonal rhabdomyosarcoma can occur as a small symptom-

less swelling (ii) the femoral neck is not an uncommon site for a metastatic tumour. It is interesting to note that only sections from the femoral tumour show some degree of differentiation, whereas sections from the organs containing the metastases reveal only the presence of undifferentiated malignant cells. The microscopic features of the malignant cells seen in the smears of the sternal bone marrow do not contribute to the diagnosis of embryonal rhabdomyosarcoma.

SUMMARY

A case of embryonal rhabdomyosarcoma is reported in which the primary is probably in the upper end of the left femur, causing a pathological fracture of the femoral neck. Clinically, the patient presented with leuco-erythroblastic anaemia, thrombocytopenia, and splenomegaly. Prolonged bleeding time led to haemorrhagic tendency and he died of cerebral haemorrhage shortly after admission.

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