CASE REPORT - MELANOMA OF THE ANORECTUM

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ABSTRACT

Melanomas of the gastrointestinal tract are rare tumours, the anorectum being the most common site. Anorectal melanomas can pose difficult diagnostic and therapeutic problems. Awareness on the part of both the clinician and histopathologist that melanomas can rarely occur in the anorectum is an important pre-requisite to its diagnosis as specific histopathological staining may be required. Therapeutically, surgery remains the primary option. Prognosis is however poor as metastatic disease is commonly established at presentation. When the tumour is sited in the rectum, the cells of origin is controversial. A case of disseminated anorectal melanoma is described.

Keywords: melanoma, anorectum

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INTRODUCTION

A case of melanoma of the anorectum is described. The clinical features of this tumour are summarised and controversies regarding the cells of origin of the tumour are highlighted.

CASE REPORT

A 55-year-old Chinese man was admitted with a history of progressive disorientation, bowel and bladder incontinence and progressive weakness of the left arm and leg over a period of 4 days. He had no other risk factors predisposing to a cerebrovascular accident apart from age.

Clinical examination confirmed a right cerebrovascular accident. Computerised axial tomographic scan of the head confirmed a large area of low attenuation in the right frontoparietal region which remained unenhanced after contrast injection, consistent with an area of infarction. There were also multiple areas of osseous destruction in the skull suggestive of metastatic deposits. Upon further examination, a mass was felt per rectally 5 cm from the anal verge. Inguinal nodes and the liver were not enlarged clinically. Flexible sigmoidoscopic examination confirmed the presence of a sessile polypoidal mass, about 4 cm in size, 5 cm from the anal verge. The lesion was not pigmented and there was no abnormal pigmentation of the surrounding mucosa. Histology revealed sheets of malignant cells with varying amounts of cytoplasm and prominent eosinophilic nucleoli. In areas, the cytoplasm contained granules of a dark pigment with staining properties of melanin. The immunoperoxidase stain was positive for S100 protein and negative for EMA (epithelial membrane antigen), cytokeratin and LCA (leucocyte common antigen).

Liver function test revealed total protein of 69g/L (62-82g/L), albumin 32g/L (37-51g/L), bilirubin 12.1umol/L (3-24umol/L), serum alkaline phosphatase(SAP) 779U/L (30-100U/L) with 46% heat stable fraction, ALT 279u/l (9-36U/L), GGT 355U/L (9-41U/L) . These suggested the presence of liver infiltration. Part of the SAP elevation could be bone derived. Ultrasonic imaging of the hepatobiliary system confirmed scattered multiple echo-poor round lesions of varying sizes throughout the liver. Some of these lesions had a "target" appearance consistent with that of metastatic deposits. Serum calcium and phosphate levels were normal. Carcino-embryonic antigen and alpha-fetoprotein were normal at 3.2ug/L (1-3.5ug/L) and 2.6ug/L (1-10ug/L) respectively. Chest X-ray did not reveal evidence of bony destruction nor of pulmonary deposits.

The patient deteriorated progressively and passed away 2 weeks post admission from the massive cerebral infarction. A trial of dexamethasone yielded no improvement.

DISCUSSION

This patient was incidentally discovered to have metastatic melanoma from an anorectal primary. The anorectum has been variously quoted as contributing from 0.4 to 3% of all melanomas[1,2] and is the third most common overall site for the tumour. It is preceded by cutaneous melanomas (75% of all melanomas) and ocular melanomas[1]. Melanomas of the anorectum are therefore rare tumours, rarer still if the tumour is sited in the rectum. It has been estimated that one melanoma occurs for every 8 squamous cell carcinoma of the anal canal and every 250 adenocarcinomas of the rectum[3]. Within the gastrointestinal tract only the oral cavity, oesophagus and gallbladder have been documented to be sites of melanomas other than the anorectum. Melanomas in the gallbladder are exceedingly rare.

This patient's tumour is sited 5 cm from the anal verge, placing it within the rectum. Definitive evidence that the primary tumour is confined within the rectum would require histological proof that the tumour is surrounded by rectal mucosa. There must also be no evidence of spread to the rectum from a primary anal focus. This data is not available as the tumour was not resected for detailed histological examination and no post mortem was performed. The siting of this tumour is best left as the "anorectum".

Melanomas sited within the rectum are interesting because their cells of origin remain controversial. Histological examinations of the anorectum confirm the rarity of melanin above the anal valves. In melanosis coli, the pigment containing cells are macrophages and the nature of this pigment remains unclear. Rectal melanomas are therefore often attributed to tu-

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mour originating and extending from anal melanocytes which have undergone malignant transformation. However, a recent report has provided convincing evidence implicating tumour originating de novo from rectal mucosal melanocytes[4].

Several published series regarding anorectal melanomas highlighted the following features of this tumour[2,5-9], namely (1) presentation is usually as a rectal mass, often mistaken for haemorrhoids. Alternatively, it may present as rectal bleeding. (2) it is a tumour of adulthood with a peak incidence at the sixth and seventh decades. There is no sex predisposition. (3) 75% of tumours are pigmented. (4) tumours are characteristically polypoidal. (5) in the case of amelanotic tumours, diagnosis rests on the application of specialised stains for melanin. The diagnosis is otherwise easily missed. Histologically, the presence of multinucleated and polymorphous giant cells are common and helpful in the identification of these tumours as they are rarely found in adenocarcinomas of the rectum nor squamous cell carcinomas of the anal canal[2]. The presence of tumour cells in clusters and prominent eosinophilic cells are further diagnostic features. Histological features could otherwise be rather non-specific and can be reported simply as anaplastic tumours. (6) 50-60% have metastatic disease at presentation. Local extension is predominantly upwards towards the rectum.

The prognosis remains dismal. Mean survival ranges from 9 to 21.5 months. The larger the primary, the worse is the prognosis. Abdominal perineal resection as compared to local excision (with or without lymph node resection) offers little improvement in survival. The actual operation chosen is therefore more dependent on technical constraints, siting of tumour and presence of local lymphatic deposits. Recurrence after apparent curative resection is common. Five-year survival for patients who have undergone potentially curative resection is quoted at 16%. The abundance of both vascular and lymphatic drainage channels for the anorectum has been cited as an explanation for the highly aggressive behaviour of these tumours. The tumour is radioresistant and chemotherapy offers little benefit.

CONCLUSIONS / RECOMMENDATIONS

Melanoma of the anorectum is a rare tumour. Given its propensity for early metastatic spread and its poor response to adjunctive therapy, prognosis is generally poor. Awareness on the part of the histopathologist and clinician that the anorectum is a site for this rare tumour is essential for its specific identification as special stains are often required to confirm the diagnosis.

REFERENCES

1. Mill SE, Cooper PH. Malignant melanomas of the digestive tract. Pathol Ann 1983; 18:2-26.
2. Wanebo HJ, Woodruff JM, Farr GH, Quan SH: Anorectal melanoma. Cancer 1981;47:1871-900.
3. Morson BC, Volkstandt H. Malignant melanomas of the anal canal. J Clin Pathol 1963; 16:126.
4. Werdin C, Limas C, Knodell R. Primary melanocarcinoma of the rectum. Evidence for origination from rectal mucosal melanocytes. Cancer 1988; 61: 1364-70.
5. Alexandra RM, Cone LA. Malignant melanoma of the rectal ampulla. Dis Colon Rectum 1977; 20:53-6.
6. Remiglo PA, Der BK, Forsberg RT. Anorectal melanoma. Dis Colon Rectum 1976; 19:350-6.
7. Chiu YS, Uyni KK, Beart RW. Malignant melanomas of the anorectum. Dis Colon Rectum 1980; 23:122-4.
8. Cooper PH, Mills SE, Allen MS. Malignant melanoma of the anus. Dis Colon Rectum 1982; 25:693-703.
9. Mason JK, Helwig EB. Anorectal melanoma. Cancer 1966; 19:39-50.