Malignant peripheral neuroepithelioma "peripheral neuroblastoma" a case report

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ABSTRACT

Peripheral neuroepithelioma is a rare malignant tumour of primitive peripheral non autonomic nervous system. The tumour can occur at any age involving soft tissue mainly of lower extremities. Other sites of occurrence are the shoulder-thoracic region and the head and neck. It is a very aggressive tumour with early metastasis mainly to the lung.

Herewith we report a case of 16 years old female who was presented with soft mass of labia `majora of 3 years duration which has been rapidly enlarging in the last 6 months. The mass was excised and followed by administration of chemotherapy and radiotherapy. The patient developed secondaries in the lung 3 months after excision, with pleural effusion, but no local recurrence. However, eight months after the diagnosis and in January 2005, the patient died from the tumour.

Keywords: Peripheral Neuroepithelioma Primitive Neuro Ectodermal Tumour "PNET" Soft tissue.

Peripheral neuroepithelioma was first described by Stout in (1918) on a tumour of the ulnar nerve composed of small rounded cells forming rosettes. In tissue culture the cells grow axons or form epithelium similar to neuroepithelium; hence the name neuroepithelioma (1).

Histologically and cytochemically, this tumour shares many features with neuroblastoma, however, there are many points of difference between the two; some of which are summarized in the table below.

CASE REPORT: The patient is 16 years female, who was presented to the surgeon with big mass in labia majora of 3 years duration with recent rapid increase in size for the last six months.

Physical examination revealed a 15x10 cm oval lobulated firm mass covered by normal skin on the right side of labium majus. There was no lymphadenopathy. The remainder of the physical examination of the patient in general, was within normal, so also were routine laboratory tests.

Ultrasound of the mass revealed solid heterogenous, echogenic tumour composed of multiple coalescent masses leading to the lobulated outline with features of necrosis. Ultrasonographic appearance of the rest of the body and pelvis were of no significance.

The tumour was noticed during surgery to be extremely vascular but well defined, so total excision was made. The post operative outcome was uneventful.

	Neuroblastoma	Peripheral Neuroepithelioma
Origin	Sympathetic nerves	- Peripheral non autonomic
Site	Adrenal gland Retroperitonium Mediastinum	Soft tissue lower extremities Soft tissue shoulder-thoracic Head and neck
Age	Children 90% <5 yr	Any age
Incidence	3rd most common in children	< 1% of all sarcomas
EM Dense core granules	(++)	(++)
VMA in urine	Elevation	Negative
Immunohistochemistry -HLA-ABC -B_2-microglobulin - S- 100 protein -Neurospecific enolase	 Negative Negative Negative Positive	 Strong positive Strong positive Negative Positive
Chromosomal translocation 11:21	Negative	Positive
Ganglionic maturation	May occur	Never occurs

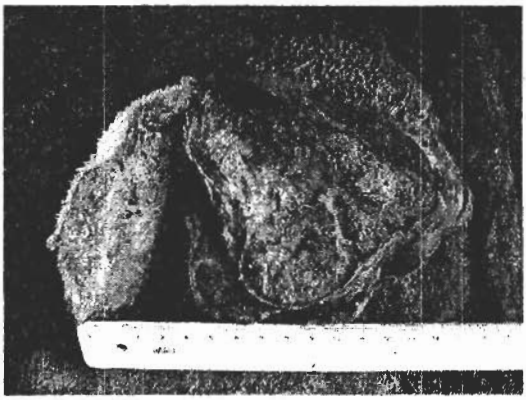

Fig. 1

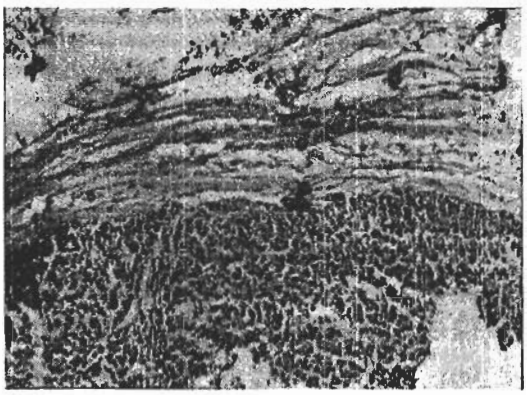

Fig. 2

One month after the operation, the patient received three courses of chemotherapy composed of adriamycin, toposid and cisplatin in addition to the radiotherapy. However at follow up, the plain roentgenograms of the chest showed secondries in the lung three months after the operation with subsequent pleural effusion, and the patient died eight months after the diagnosis.

Pathology: Gross examination (Fig.1): The mass was firm in consistency and lobulated 15x10 cm in size, covered by skin which can easily be mobilized over the tumour. Slicing revealed dark brown and solid cut surface with wide areas of necrosis.

Histopathology: The tumour was very cellular (Fig.2), with extensive necrosis and cellular viability around vessels. The cells are small and rounded arranged all over in Homer-Wright rosettes (Fig.3). Vascular invasion was noticed at the periphery of the tumour (Fig.4) with retained rosettes formation of the intravascular clumps.

DISCUSSION

Malignant peripheral neuroepithelioma is a rare tumour of peripheral nerves which has been sporadically reported in various sites but mainly in soft tissue of lower extremities[1,2], head and neck[3] and scapular thoracic region[2,3]. Histologically, the tumour is composed of small round cells with Homer-Wright rosettes which makes it very reminiscent to neuroblastoma. However, the two tumours vary in clinical aspect, behaviour, prognosis, and response to treatment[1]. In our case, the patient had the tumour at the upper medial site of the right thigh which extended to the labium majus and presented as such. Local excision, radiotherapy and chemotherapy had little effect as the patient developed pulmonary metastasis with pleural effusion only three months afterward; however there was no local recurrence.

Malignant peripheral neuroepithelioma has been reported in the literature under varieties of names including peripheral neuroblastoma[1] and Askin's tumour[2]. Recently, in a report of five cases at the head and neck, Nikitakis et al,[3] included the tumour within the larger family of peripheral neuroectodermal tumours (PNET).These tumours share the characters of being all formed of small round cells with variable degree of rosette formation and with intracytoplasmic dense core granules. Included under this family are three main groups:

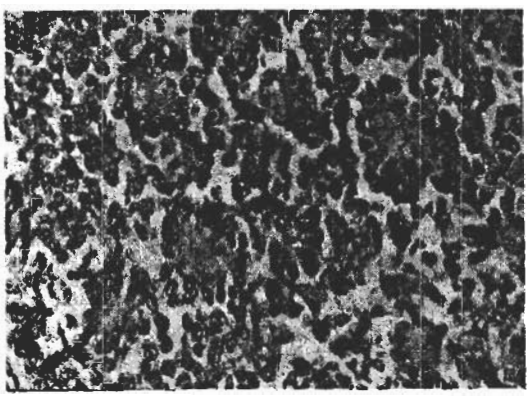

Fig. 3

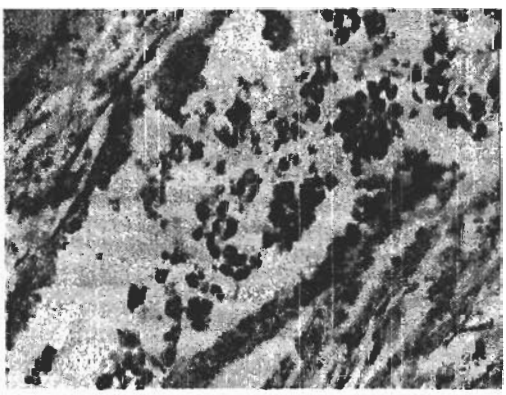

Fig. 4

1-Central (PNET) tumour of the C.N.S like medulloblastoma.
2-Neuroblastoma of sympathetic nerves.
3-Peripheral neuroepithelioma which include tumours of non-autonomic nerves and Ewing's sarcoma of osseous and extraosseous types (3).

Accordingly, the tumour has been renamed as malignant PNET; peripheral neuroepithelioma; is a rather cumbersome name for such a rare and unusual tumour.

The behaviour of this tumour has been described as poor and dismal in all available reports[1-4] with 3 years survival of less than 50% and early metastasis to lung, liver and bone. However, factors which may alleviate the prognosis are radical excision with clear margins, radiation before surgery, combination chemotherapy as well as indication of the site of the primary tumour[3]. Our patient died only eight months after the diagnosis.

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