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Ewing's sarcoma of the retropharynx

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Abstract
Ewing's sarcoma arising in the head and neck region is very rare. A case arising from C2 in a young child is presented. The unusual presentation, differential diagnosis and pathological features are discussed.

Key words: Sarcoma, Ewing's; Pharynx; Cervical vertebrae

Introduction

Ewing's sarcoma is an uncommon malignant neoplasm, which is locally highly aggressive and generally occurs in the first three decades of life more commonly in males (Ferlito, 1978). The most common sites of occurrence are the long bones. In otolaryngological practice this tumour is very rare but may involve the mandible, cervical vertebrae, or temporal bone.

Case report

A 4-year-old child was admitted to Birmingham Children's Hospital for investigation of a persistent sore throat, cervical lymphadenopathy and a painful stiff neck. Prior to admission he had been treated with repeated courses of antibiotics on the basis of a diagnosis of tonsillitis. Physical signs included a mild pyrexia of 37.5°C, unremarkable tonsils but the left tonsil was displaced anteriorly. There was an enlarged, tender left jugulodigastric lymph node measuring 2 × 2 cm associated with numerous other small, nontender cervical nodes and a stiff painful neck.

Full blood count, alkaline phosphatase and erythrocyte sedimentation rate were all normal. Blood cultures, antibodies to Epstein-Barr virus and bone marrow examination were all negative. There was no excess catecholamine in the urine. Plain films of the cervical spine (Figure 1) showed the body and arch of C2 appeared dense with loss of normal cortico-medullary differentiation. A CT scan (Figure 2) showed a large pre-vertebral mass running from C1–C4 with a large intraspinal extent compressing and displacing the spinal cord to the right. An isotope bone scan (Figure 3) showed increased uptake in C2 but no other abnormality.

Pathological findings

A biopsy obtained from the retropharyngeal mass showed a neoplasm consisting of small dark round cells with scanty basophilic cytoplasm and dense nuclei (Figure 4). Occasional pseudo-rosettes were seen but there were no Homer-Wright rosettes. The reticulin pattern showed condensation around blood vessels with minimal intercellular reticulin fibres. There was both intra- and extracellular glycogen. Immunohistochemistry showed strong cytoplasmic postivity for vimentin (a mesenchymal intermediate filament). All other immunostaining, including neural markers (NSE, PGP 9.5 and S100) a muscle marker (desmin) and a pan leukocyte marker, (LCA which recognizes CD45) were negative.

Ultrastructurally the tumour cells contained intracytoplasmic glycogen. Tight junctions and occasional intermediate filaments were seen (Figure 5).

A diagnosis of Ewing's sarcoma was made and the child was commenced on cyclical chemotherapy: ifosamide. Mesna, vincristine, actinomycin D and adriamicin. He also received a course of radiotherapy to the tumour mass.

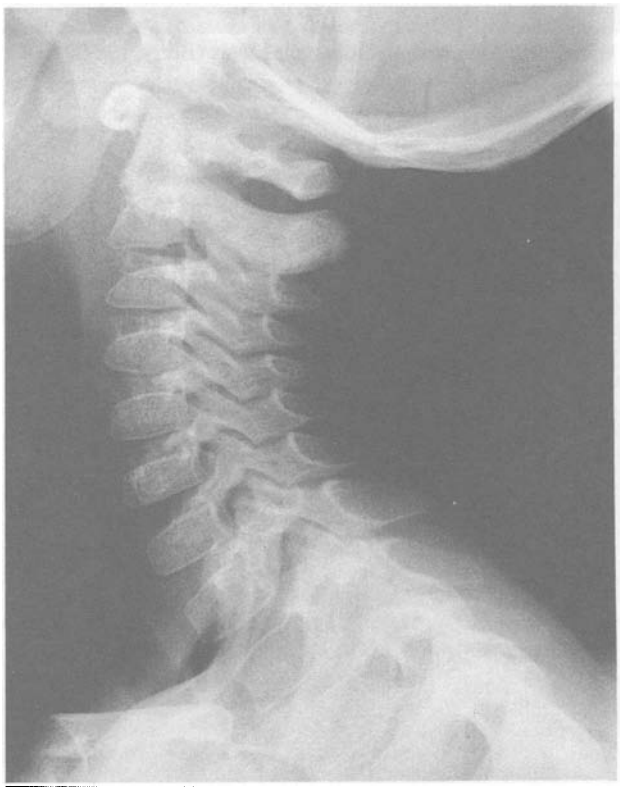

FIG. 1

Plain film, lateral view of the cervical spine showing density of the arch of C2 with loss of normal cortico-medullary differentiation.

From the Birmingham Children's Hospital, Birmingham.
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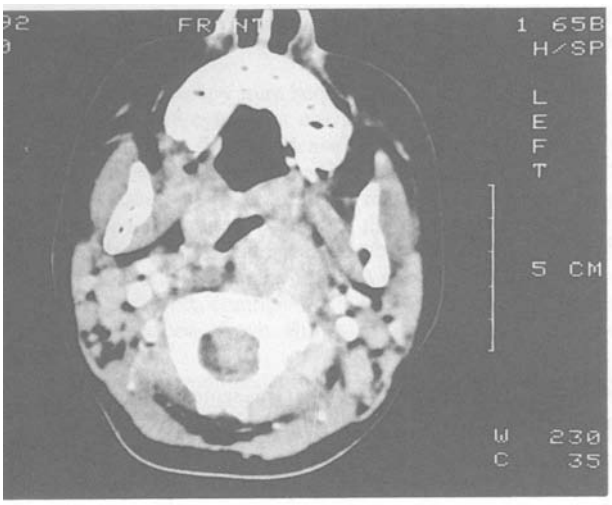

FIG. 2

CT scan showing a large pre-vertebral mass with an intra- and extraspinal component.

Discussion

This tumour is one of the varieties of small round cell malignant tumours of childhood. The differential diagnosis of this group in childhood is Ewing's sarcoma, a primitive neuroectodermal tumour (PNET), embryonal rhabdomyosarcoma, neuroblastoma and non-Hodgkin's lymphoma (Triche, 1982). A distinction between these entities may be difficult as all may present as undifferentiated round cell tumours in bone, soft tissue or lymph nodes and be indistinguishable by light microscopy. Special techniques such as electron microscopy, histochemistry, immunohistochemistry and tissue culture may be required to make a definitive diagnosis (Berry, 1987).

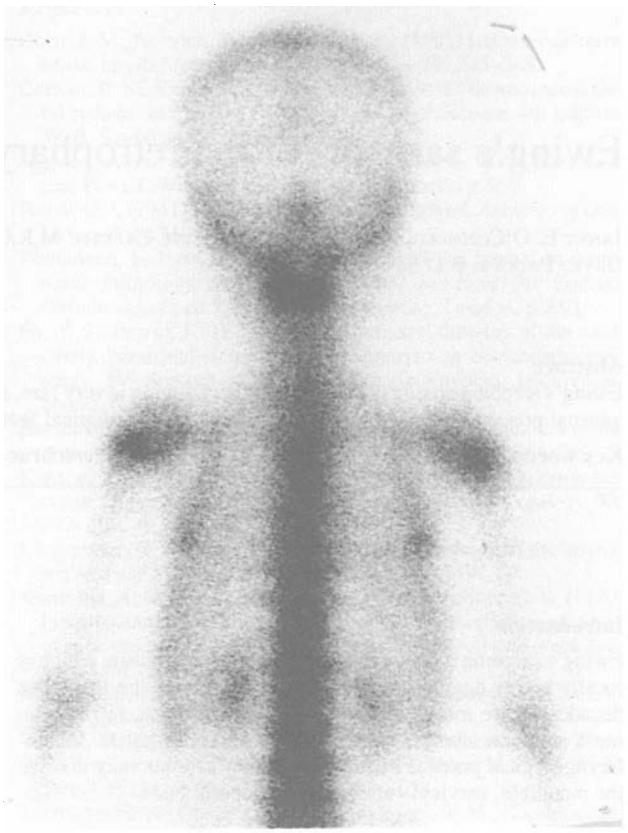

FIG. 3

Isotope bone scan showing increased uptake in C2.

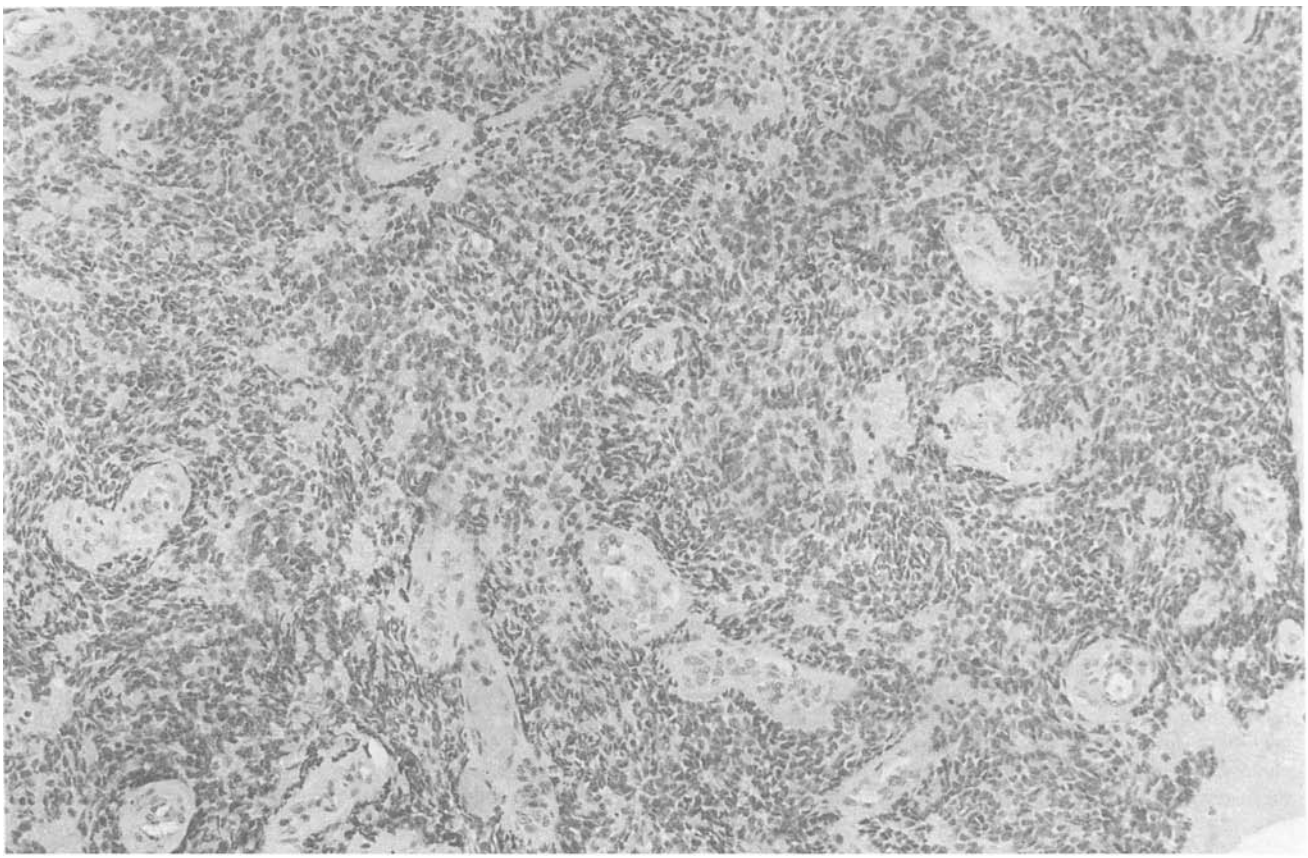

FIG. 4

Light microscopy. Sheets of tumour cells with uniform appearance. (H & E × 190).

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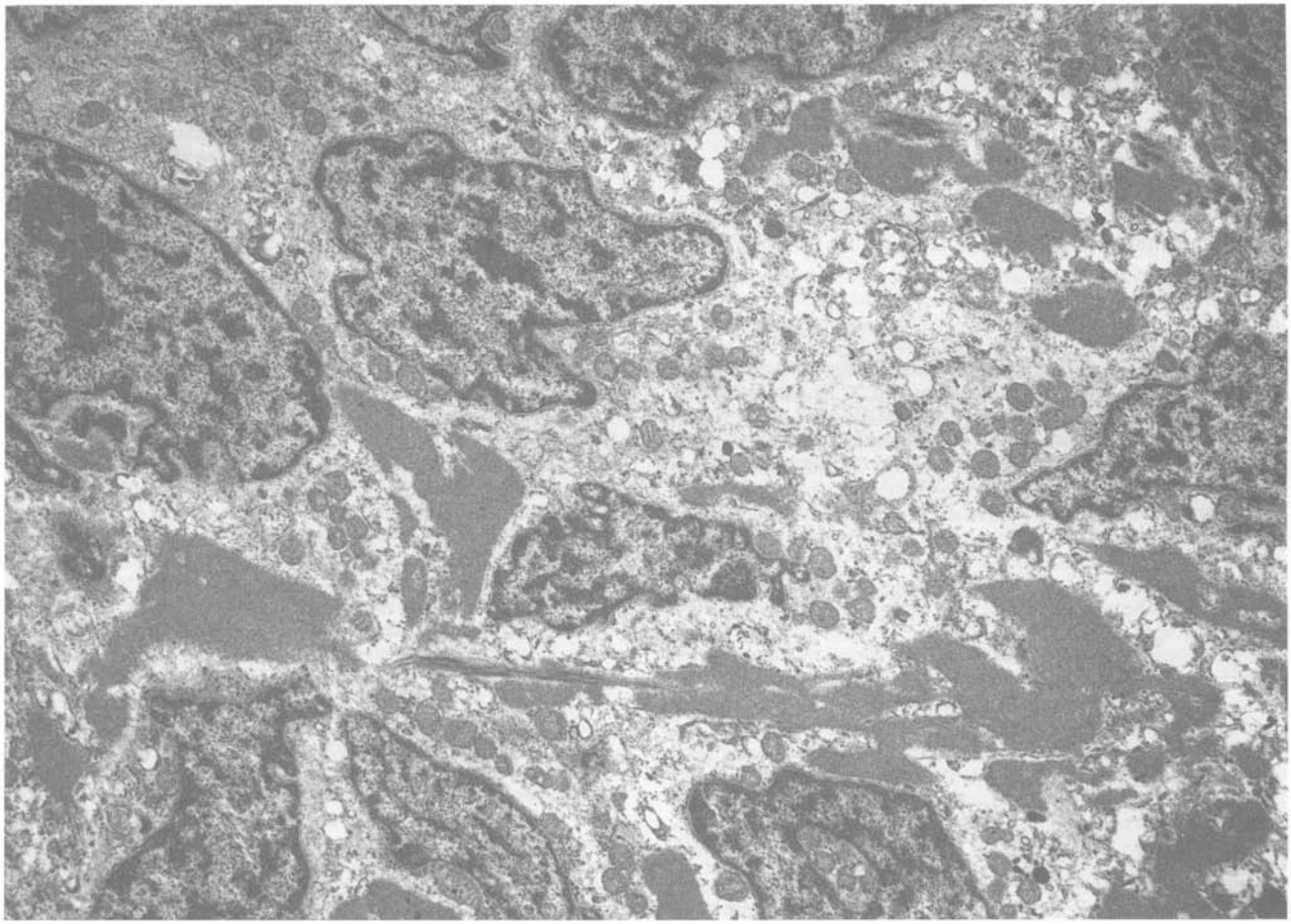

FIG. 5

Electron microscopy appearance showing tumour cells containing irregular nuclei with margination of chromatin, rare intracytoplasmic organelles and occasional mitochondria. (× 3300).

The tumour cells contain glycogen and this makes diagnosis of neuroblastoma or non-Hodgkin's lymphoma unlikely. Furthermore neuroblastomas are neurons specific enolase (NSE) positive and the majority have consistently raised levels of urinary catecholamines (Berry, 1987). Of the tumours that characteristically contain glycogen, rhabdomyosarcoma can be excluded by lack of expression of myoglobin and desmin. It is true, however, that some cases of rhabdomyosarcoma, if poorly differentiated, can be desmin/myoglobin negative. Here electron microscopy can be helpful in demonstrating myofilaments.

In this case the differential diagnosis was between primitive neuroectodermal tumour (PNET) and Ewing's sarcoma. These two tumours appear to be closely related. PNET of bone is similar to Ewing's sarcoma but shows evidence of neuroectodermal differentiation, seen as NSE positivity of tumour cells and the formation of true Homer-Wright rosettes (Jaffe *et al.*, 1984).

The histiogenesis of Ewing's sarcoma remains unclear. Ewing's original suggestion of endothelial cell origin has not been substantiated. There are two other possibilities: an origin from primitive mesenchymal cells; or the neural crest (Berry, 1987).

Ewing's sarcoma is an aggressive tumour most commonly affecting the long bones of children aged between 10 and 15 years. It comprises four to six per cent of all primary bone tumours: one to four per cent of all cases of Ewing's sarcoma affect the head and neck and in this area the skull or mandible is the most frequently affected region (Siegal *et al.*, 1987). Siegal *et al.* (1987) for the Intergroup Ewing's Sarcoma Study Group collected 805 cases between 1972 and 1987. Of these cases only 32 had Ewing's sarcoma of the head and neck and there were seven cases only arising in the cervical vertebrae, all older than our case.

The commonest presenting features are usually a mass or swelling at the site of the tumour. Localized pain and tenderness is often a prominent feature. Neurological and ocular symptoms are also common. Systemic signs, fever, leucocytosis and a raised erythrocyte sedimentation rate also occur and are thought to be associated with a poorer prognosis.

Radiologically, Ewing's sarcoma may be confused with many other benign and malignant conditions of bone. The commoner radiological findings are those of permeative (mottled osteolytic) destruction of bone with cortical expansion and soft tissue swelling. The periosteal reaction may be lamellated or spiculated. In the head and neck the typical 'onion peel' periosteal reaction is often absent (Sneige and Batsakis, 1989).

The prognosis for Ewing's sarcoma has improved greatly with treatment utilizing radiotherapy to the primary site and combination chemotherapy.

Siegal *et al.* (1987) for the Intergroup Ewing's Sarcoma Study group found that patients who had biopsy only or complete surgical excision (followed by radiotherapy and chemotherapy) survived longer than those who had incomplete excision. Eighty per cent of their patients with Ewing's sarcoma of the head and neck had survived three years. Prior to 1975 of 1000 patients with Ewing's sarcoma reported in the world literature and treated with radical surgery or radiotherapy only about 100 survived five years or longer (Sneige and Batsakis, 1989).

Conclusions

Our case was unusual with respect to site, young age and superficial presentation as a common acute inflammatory con-

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dition, with fever, tender cervical lymphadenopathy and a retropharyngeal mass. Precise pathological differentiation distinguishing it from the other small round cell tumours of childhood is important for correct treatment regimes and prognosis.

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