Alveolar soft part sarcoma: a case report

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ALVEOLAR SOFT PART SARCOMA is an uncommon clinical entity, the pathogenesis of which has been controversial. Recently, Dutt et. al. (1969) have reported one such case from Malaysia. The following case report throws further light on the nature of the tumour.

Case Report

A healthy 42-year-old Chinese woman was admitted to the University Hospital on 6 June 1969. She had noticed a pea-size lump growing on her right arm for the last four years. At first, the lump very slowly increased in size and asymptomatic. One month prior to admission, the rate of growth had acclerated and the lump become painful.

On examination, a rounded firm swelling measuring 4 cm. across was found on the postero-medial aspect of her right arm. It was fairly mobile but appeared to tether to the tendon of triceps. The overlying skin was free and unremarkable and the movements of the right elbow were unrestricted.

Investigations showed a normal blood picture with the erythrocyte sedimentation rate 10 mm. per hour. Roentgenogram of the chest was normal while that of the elbow showed a normal bony architecture with a shadow of a soft tissue mass 5 cm. proximal to the joint (Fig. 1). A provisional diagnosis of solitary neurilemmoma was made and on 13 June 1969, the tumour was excised under general anaesthesia. At operation, the tumour was found to be attached to the triceps tendon but was well capsulated. The cut surface of the tumour appeared firm greyish-yellow tissue with foci of haemorrhage.

Histology

Sections showed areas of cluster of cells surrounded by connective tissue septa containing fine vascular channels. An organoid pattern was thus produced (Fig. 2). The structural units appeared alveolar and in areas appeared as solidly growing cell aggregates. Reticulum stain did not show the isolation of individual cells by fibres, but each unit was seen to be surrounded by a network of collagenous connective tissue septa. The tumour cells varied in size and normally contained one or two vesicular nuclei, although as many as four were observed within some cells. The nuclei possessed prominent nucleoli. Mitotic figures were rare. The cytoplasm in haematoxylin and eosin preparations was characteristically eosinophilic and finely granular (Fig. 3). No striations or fibrils were seen. Diastase resistant PAS positive crystals were present within the cytoplasm of the tumour cells (Fig. 4). The diagnosis of alveolar soft part sarcoma is supported by the morphology of the tumour cells, the alveolar pattern and the presence of diastase resistant PAS positive crystals in many of the tumour cells.

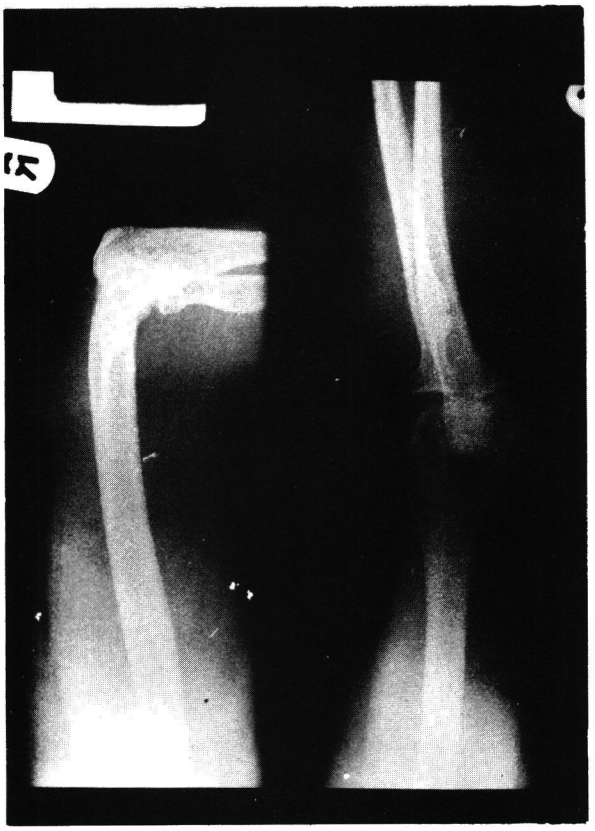

Fig. 1

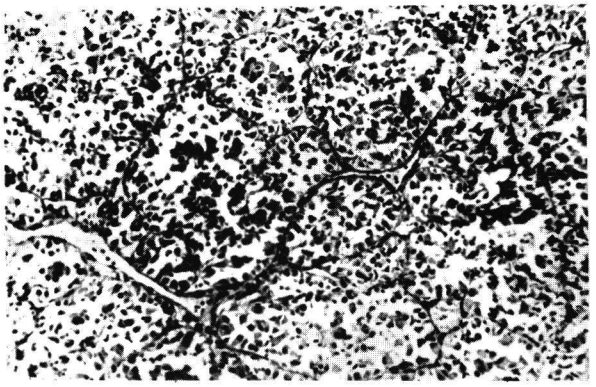

Fig. 2

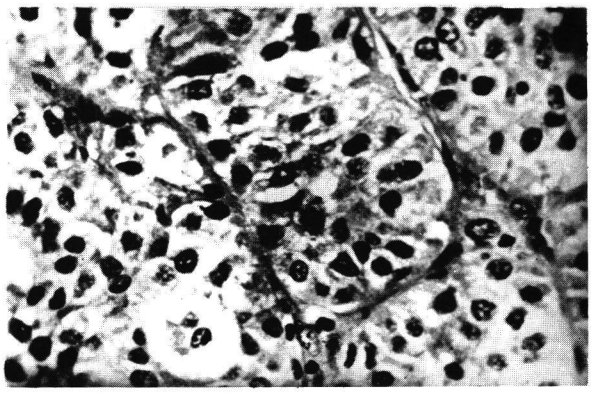

Fig. 3

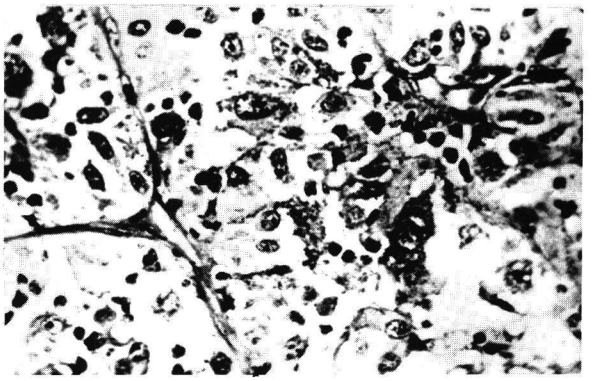

Fig. 4

Discussion

Christopherson, Foote and Stewart in 1952 described morphologically identical growths of unknown classification which they found amongst the undiagnosed tumours at the New York Registry of Cancer. Their clinical and histological similarity were remarkable. Subsequently, many such tumours have been documented (Farquharson 1960, Hicks and Leitch 1955, Mcfarlane and McGregor 1958 and Randall and Walter 1954). These tumours occur predominantly in women and are commonest in the late teens or early twenties. They are invariably related to skeletal muscles and are slow-growing and asymptomatic until late. Though well capsulated, the tumour is malignant and often metastasises to the lungs. Microscopically, the appearances of the tumour are characteristic, being uniform both at the primary and metastatic sites. Large eosinophilic cells, with alveolar or pseudo-alveolar arrangements, lie in close relation to endotheliúm-lined blood vessels. Periodic acid stain shows abundant fuchsinophilic cytoplasmic granules. In the absence of a better name, Christopherson (1952) called these tumours alveolar soft part sarcoma. They can be easily mistaken for metastatic adenocarcinoma, endothelioma, liposarcoma, myosarcoma and myoblastic myomas. However, their organoid structure and absence of neurofibrils or cross striations are usually distinctive.

The pathogenesis of this rare but very interesting tumour is not known. Swetana and Scott in 1951 described eight tumours with nearly identical histological pattern and from their resemblance to carotid body-like tumours, thought them to be malignant, non-chromaffin paragangliomas. Similar views have been expressed by Barbera and Fiore-Donati (1953), Randall and Walter (1954), Willis (1967) and Evans (1966). Karnauchow and Magner (1963) also noted the overlapping histological patterns and suggested that the alveolar soft part sarcomas arise from undifferentiated primitive neural crest cells, thus explaining the presence of such tumours in places where paragangliomas are anatomically non-existent. Fisher (1956) thought this tumour of neural origin, as biochemical methods show that the cytoplasmic granules of the tumour cells are mainly cerebrosides with a small amount of polysaccharides. The similarity of the cytoplasmic granules of alveolar soft part sarcoma to those of malignant granular cell myoblastoma and the myelin of peripheral nerve, leaves no doubt as to their neural origin. Christopherson and his co-workers (1952), in their original paper, found impressive resemblance of alveolar soft part sarcomas to malignant myoblastomas. MacFarlane (1958) does not believe in the separate existence of the alveolar soft part sarcoma or malignant myobiastomas. The close proximity to muscles, granularity of the cytoplasm and distinctive organoid histological pattern of both these tumours are, according to him, typical of non-chromaffin paraganglioma.

However, recently Shipley et. al. (1964) have shown, with the help of the electron microscope, definite and specific crystalline structures in the intracytoplasmic granules in alveolar soft part sarcoma, which are not seen in myoblastoma or carotid-body tumour. The presence of these characteristic crystalline structures are a definite diagnostic aid. Udekwu and Pulvertaft (1965), in tissue culture studies of a typical case of alveolar soft part sarcoma in a young Nigerian male, found that the behaviour and the characteristics of these tumour cells contrasted sharply with tissue culture of carotid-body tumours as reported by Costero (1963). They conclude that the alveolar soft part sarcoma is not a paraganglioma.

Summary

A rare case of alveolar soft part sarcoma in a Chinese female is described. The controversy regarding its histogenesis is reviewed.

Acknowledgement

The histological sections of this case was sent to the Armed Forces Institute of Pathology, Washington D.C., U.S.A. for consultation and they concurred with the diagnosis.

We are indebted to Dr. Ferguson of the Department of Pathology for reviewing the paper; the Department of Radiology for the roentgenograms; the Department of Medical Illustrations for the micro-photographs; and to Professor J.F. Silva, Department of Orthopaedic Surgery, for permission to publish the case report.

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