Leading Article

Malignant fibrous histiocytoma

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Malignant fibrous histiocytoma (MFH) was the name proposed for a group of soft tissue sarcomas on the basis of tissue culture studies which suggested the tumour cells were of histiocytic origin. These sarcomas had been previously classified as poorly differentiated sarcomas of other types (particularly fibrosarcoma). As new histological criteria were established for both MFH and fibrosarcoma, many sarcomas were diagnosed as MFH with the result that MFH is now considered the commonest soft tissue sarcoma. Further work, largely by Enzinger and his co-workers at the Armed Forces Institute of Pathology, introduced the sub-types of soft tissue MFH that are recognized today—pleomorphic-storiform, myxoid, inflammatory, giant cell-rich and angiomatoid. The histopathological features of the various sub-types have been well illustrated by Enzinger and Weiss.

The spectrum of fibrous histiocytic tumours was expanded to include benign tumours of dermis and tendon sheath, such as dermatofibroma and benign synovioma, and soft tissue tumours of intermediate malignancy such as dermatofibrosarcoma protuberans. Bone tumours with a similar histological appearance were included as benign and malignant fibrous histiocytoma of bone. With increasing recognition by pathologists of the histological pattern of MFH, tumours with this appearance were diagnosed as MFH at more and more sites other than the soft tissues. Two further cases of MFH at uncommon sites are reported in this issue.

Grieco and his colleagues report a case of a 42-year-old man with a 3-month history of pyrexia who was found to have a MFH of the sternum. The tumour was deemed inoperable and was treated with chemotherapy. Residual tumour could not be found in the sternum at autopsy. MFH is a rare bone tumour; for example, it represents only 0.8% of the 6514 primary bone tumours described from the Mayo Clinic. Sixty-three percent of tumours involve the long bones and the sternal location of the reported case is most unusual. Like Grieco’s case, the majority of osseous MFHs are of the pleomorphic-storiform type. The optimal treatment for these tumours is chemotherapy followed by surgical excision and, for long bone tumours, prosthetic replacement.

Aggarwal and colleagues describe a MFH of the posterior mediastinum in a 16-year-old girl who presented with pyrexia. The tumour was surgically excised and the pyrexia subsided post-operatively. The mediastinum is another rare site for MFH. Since the initial description of two mediastinal cases by Mills et al., only single cases have been reported.

A further unusual feature of the two cases of MFH in this issue is the prominent pyrexia. Many neoplasms are associated with pyrexia, notably gastric, pancreatic, hepatic and renal carcinomas and leukaemias and lymphomas. Most cases are due to associated infections or the products of tissue injury. Pyrexia in association with MFH has been noted previously and was present in 2 out of 200 cases of soft tissue MFH. Other occasional associations with MFH have included episodic hypoglycaemia and rapid enlargement of the tumour during pregnancy.

Despite the evident attractiveness to pathologists of fibrous histiocytoma as a diagnosis, cracks are beginning to appear in the concept of fibrous histiocytic tumours. The diagnosis of pleomorphic MFH (the commonest sub-type) in particular is coming under strong attack with the question being posed whether it represents a real biological entity or has it become a ‘diagnostic dustbin’ like the poorly differentiated fibrosarcoma of old.

Doubt has been cast on the original tissue culture studies and it now seems probable that the tumours studied were malignant lymphomas. The finding that some cells within MFH stained with the first ‘histiocytic’ markers used in immunohistochemistry—polyclonal antisera against alpha-1 antitrypsin, alpha-1 antichymotrypsin and lysozyme had been taken to support histiocytic differentiation but this support has been eroded by subsequent studies showing these enzymes to be rather ubiquitous and by studies using newer monoclonal antibodies (with greater macrophage specificity) revealing that only a very small number of true histiocytes are present within...
these tumours—cells thought to be a transient macrophage population as can be seen in many tumours. That fibrous histiocytomas do not contain neoplastic histiocytes may be unsatisfactory to terminological purists, but it does not matter in any practical sense—what’s in a name? and our concepts of tumour histogenesis from, rather than towards, cell type may be at fault.

The accusations being levelled at pleomorphic MFH are more serious. That the histological pattern of pleomorphic MFH can be seen in the dedifferentiated parts of a number of tumours has been recognized for some years. What then, if tumours currently diagnosed as pleomorphic MFH are more thoroughly examined? By sampling the tumour even more widely than has been current practice, by using a panel of immunohistochemical markers and by submitting a portion of each tumour for electron microscopy, would evidence be found to make a diagnosis of another type of tumour? Some pathologists assert this to be so for many pleomorphic MFHs of soft tissues but most pathologists find that, even after such procedures, there remain some tumours for which pleomorphic MFH is the only creditable diagnosis.

As treatment for sarcomas becomes increasingly sophisticated, this is important; for only by knowing what exactly is being treated can we hope to improve outcome. It may be that sarcomas consisting mostly of MFH-like tumour will behave in the same biological manner but, at present, we do not really know. Because of the rarity of these tumours at some sites, case reports such as those of Greico et al. and Aggarwal et al. will continue to have their place.

References


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doi: 10.1136/pgmj.65.770.872

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