Malignant Fibrous Histiocytoma of Ethmoid Sinus

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Malignant Fibrous Histiocytoma (MFH) is a rare tumour to involve ethmoid sinuses. So far only 7 cases have been reported in the literature to our best of knowledge. The disease predominantly affected younger people with no significant sex predilection. The aggressive behaviour of the tumour clinically is an important parameter to clinch the malignant nature of the disease. Wide surgical resection is the accepted method of treatment. Local recurrences are frequent and distant metastasis are not known from ethmoid sinus.

INTRODUCTION

Malignant fibrous Histiocytoma (MFH) is an unique tumour with aggressive clinical behaviour and varied histological appearances. The tumour inspite of its clinical invasiveness could be benign under the microscope. It is extremely rare to involve the paranasal sinuses. We report a case of MFH of ethmoid with orbital extension in a young girl.

CASE REPORT

K.B., a 12-years old girl presented on 11.4.91 with the complaints of swelling left medial canthus, nasal obstruction and proptosis of 6 months duration. Before being referred to our department she underwent exploration of the lesion elsewhere but only a biopsy was taken which proved to be inconclusive. On examination she had fullness of the medial canthus on left side with an incision scar. The left eye was pushed forward and laterally with no restriction of movements (Fig.1). The vision was normal. Anterior rhinoscopic examination showed obliteration of left middle meatus and

the middle turbinate pressed firmly against the nasal septum. CT Scan evaluation of para nasal sinuses showed extensive bone destruction of left ethmoidal labyrinth with invasion of orbit and

Fig.1

Clinical photographs showing the displacement of the eye

maxillary antrum. The expansions are upto septum medially and inferior turbinate inferiorly, with superior bowing of the roof of ethmoid sinus and floor of anterior cranial fossa (Fig.2). The tumour was resected radically using an Extended Lateral Rhinotomy approach (Fig.3). There was destruction of nasal bone, frontal process of maxilla, lacrimal bone and lamina-paparea on left side. The superior extent was upto the skull base of anterior cranial fossa and cribriform plate medially. The large cavity was made to drain through the nose and the patient had an uneventful recovery.

The microscopic examination of the specimen showed short intersecting fascicles of fibroblasts

arranged in storiform pattern with few histiocytes. Lymphocytes are also seen scattered in a delicate

and with the addition of our case it appears that younger age group are more effected with no sex predeliction in ethmoidal sinus disease.

Fig.2

CT Scan Coronal View of PNS showing the Extensions of the Tumour

collagen network (Fig.4). A diagnosis of MFH was put forward also taking into consideration the clinical aggressiveness of the tumour.

DISCUSSION

MFH frequently occurs in limbs and retroperitoneal structures. About 3% of all reported cases have been found in head and neck. Involvement of paranasal sinuses is extremely rare. So far only seven cases have been reported to arise in the ethmoids, since the first description of the case by Townsend et al in 1973. The ethmoid cells may secondarily be involved by the tumour extension from the adjacent sites (Rice et al 1974, Spector and Ogura 1974, Perzin and Fu 1980). There are sporadic case reports of MFH arising from the frontal sinuses (Schaefer et al 1980) and maxillary sinus (Mugliston and Shaw 1984). While reporting a case (Hazarika et al 1985) have made a detailed analysis of all the so far reported cases arising from the ethmoid sinus. The age group ranged from 3 years to 51 years

Fig.3

Resected Specimen

The tumour arises from a primitive mesenchymal cell or a histiocyte and composed of both fibrous

Fig.4

Photo Micrograph of Fibroblasts arranged in storiform pattern with few Histiocytes

tissue and histiocytes. The tumour is classified as fibrous xanthoma when there is predominance of fibrous tissue over cells with intracellular lipids and only a few histiocytes. It is labelled as histiocytoma if it is mainly made of histiocytes. The histological diagnosis is possible after

observing the spindle cells and fibre bundles arranged at right angles to one another along with the histiocytes. Tumours like rhabdomyosarcoma, fibrosarcoma may have to be entertained in the differential diagnosis.

The unique features of the tumour is despite being benign under the microscope, clinically it shows aggressive behaviour. Apart from this it has been shown that there are no definite histologic criteria for malignancy. For most of the tumours determination of malignancy by histologic findings alone may be difficult or impossible (Townsend et al 1973). Electron microscopic studies have also failed to establish valid ultrastructural criteria for malignancy (Brookes & Rose 1983). The aggressive behaviour of the tumour is also an important criteria in confirming the malignant nature of the tumour (Ogura et al

1980, Brookes & Rose 1983). The rapidity of the growth within 6 months with bone destruction and invasion is an indication of malignant nature of the tumour in our present case.

Wide surgical resection is the accepted method of treatment in cases of MFH involving the ethmoid sinus. No added benefit has been noted when combined with pre or post operative radiotherapy and with chemotherapy. However, reported cases are only very few to draw conclusion on the best line of management of MFH involving the ethmoid sinus. Local recurrences are frequent. Out of 7 cases of ethmoidal MFH, 5 cases had local recurrence ranging from 5 months to 9 years. The present case is free of any recurrence after a period of 3 years as evident by regular follow up. Distant metastasis have not been reported in MFH involving the ethmoid sinus.

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