ENDOSCOPIC RESECTION OF A RARE CASE OF SOLITARY FIBROUS TUMOUR OF THE NOSE AND PARANASAL SINUSES

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ABSTRACT: A solitary fibrous tumour is a rare neoplasm of mesenchymal origin and arises mainly in the pleura. It is characterised by spindle cells, branching blood vessels, stromal hyalinization and CD34, 99 immunoreactivity. Occurrence in the nasal cavity is extremely rare. We report a case of nasal solitary fibrous tumour in a 66 year old lady who presented with right sided nasal obstruction, headache and profuse nasal bleeding of 3 months duration. Mass was seen arising from the right middle turbinate with extension into the right middle meatus, ethmoid sinus and sphenoid sinusoidal recess. The tumour measuring 3.2x1.5x3.5 cm was successfully resected by endoscopic surgery. There has been no evidence of tumour recurrence in the 24 months follow-up following surgery.

KEYWORDS: Solitary Fibrous Tumour; Endoscopic Resection; Nasal Cavity; Epistaxis.

INTRODUCTION: CASE REPORT: A 66 year old female patient presented with complaints of right sided nasal obstruction which was gradually progressive since 3 months. It was associated with intermittent profuse right nasal bleeding and headache.

Diagnostic nasal endoscopy revealed a pinkish lobulated mass arising from the right middle meatus and anterior end of the right middle turbinate. It was cystic in consistency and bled on probing. There were haemorrhagic spots at the attachment of right middle turbinate. No postnasal extension was seen. Biopsy was abandoned due to heavy bleeding.

Otolaryngoscopic examination was within normal limits. Contrast enhanced CT scan showed an oval enhancing lesion measuring 3.2-1.5-3.5 cm occupying right side of ethmoidal sinus and projecting into the right nasal cavity closely abutting turbinates and septum in the right nasal cavity. There was no evidence of adjacent bony erosion (Fig. 1, Fig. 2).

Patient underwent endoscopic excision of mass from right nasal cavity under general anaesthesia. Base of mass attached to the anterior end of middle turbinate was cauterised and removed enblock. Undecnecotomy was done. Mass was seen extending to the anterior and posterior ethmoid sinus and sphenoid sinusoidal recess on the right side. Mass was removed in toto and complete exteration of all ethmoid air cells were done. Haemostasis was achieved.

Histopathological examination revealed section showing a neoplasm composed of spindle cells arranged in sheets and bundles with areas consisting of sclerosing bands of collagen and thin vascular spaces. Cells were seen with scanty eosinophilic cytoplasm and elongated dark nuclei. Areas with inflammatory infiltration and haemorrhage were seen. There was no evidence of mitosis or necrosis. Impression was suggestive of solitary fibrous tumour (Fig 3).

Immunohistochemistry was consistent with solitary fibrous tumour with CD 99 positive and CD34 positive.

Post-operative endoscopic suction clearance was done to clear blood clots and secretions. Follow-up endoscopy revealed a healthy right ethmoidal cavity with no recurrence. Follow-up of 24 months following surgery showed no tumour recurrence.

DISCUSSION: Solitary or localised fibrous tumour is an uncommon neoplasm that commonly arises from the pleura. It was first described as a distinct neoplasm in 1931 by Klemperer and Rabin who distinguished it from the more prevalent and aggressive diffuse malignant mesothelioma. These tumors are usually round to oval and often attached to pleura by a vascular pedicle. They have been described throughout adulthood and occur equally in men and women.

In the head and neck, this tumour may involve the orbit, salivary glands, soft tissues, eyelids, mouth and nose, nasopharynx, oropharynx, and the thyroid gland.

The diagnosis of this tumour may be difficult when it is not located in the pleura, given the variability of its histology. Typical microscopic findings are: storiform growth pattern, fusiform cells with no atypias, alternating dense cell and hypocellular areas and prominent branched vascularization similar to that of hemangiopericytomas. Primary histopathologic differential diagnostic considerations include angiofibroma, fibrous histiocytoma, spindle cell carcinoma,
spindle cell melanoma, fibromatosis, fibrosarcoma and fibroosseous lesion such as ossifying fibroma.

Immunohistochemistry is a useful adjunct for differentiating other neoplasms.\(^{(3,4,5)}\) Histological studies in Solitary fibrous tumours diffusely express vimentin and CD34 protein, Bcl-2 and CD 99 focally and are negative for muscle and epithelial cell markers.\(^{(5,6)}\) Demonstration of CD34 reactivity is a positive immunophenotype marker in 79% of Solitary fibrous tumours. It is useful for distinguishing fibrosarcoma from Solitary fibrous tumours.

Nasal Solitary Fibrous tumours usually result in progressive nasal obstruction, occasionally epistaxis, rhinorrhoea, anosmia, headache, facial pain and visual disorders due to compression of the orbit.\(^{(3)}\) No particular area of involvement of the nose or sinuses has been identified as occurring more frequently, although this would be difficult to discern with the limited number of cases described.

Based on symptoms and radiological findings the clinical differential diagnosis of nasal cavity Solitary fibrous tumours should be made with fibrosarcoma, hemangiopericytoma and nasopharyngeal carcinoma.

The endoscopic appearance is routinely described as that of a firm, lobulated and well encapsulated mass without surrounding tissue reaction. The differential diagnosis of lesions with this clinical appearance is very broad and includes epithelial neoplasm, esthesioneuroblastoma, meningioma, hemangiopericytoma, lymphoma, schwannoma, leiomyoma, angiofibroma, fibromatosis, malignant fibrous histiocytoma and fibrosarcoma.

CT findings are that of a smooth expansile soft tissue mass with surrounding bony remodeling or erosion from pressure effect. Solitary fibrous tumours show some characteristic magnetic resonance imaging findings including a well circumscribed solid mass that is hypo to isointense on T1 weighted images and has a prominent but heterogeneous enhancement with gadolinium. In addition variable hypo or more hyperintensity on T2-weighted images is also seen.\(^{(6,7)}\) Full surgical removal is curative in most cases.\(^{(8)}\)

The predominantly benign nature of nasal and extrapleural Solitary fibrous tumours contrast with the more aggressive behaviour found in 23% of pleural tumours.\(^{(3)}\) The prognosis may be based on the presence or absence of histological findings of malignancy, such as significant cellularity, a high mitotic index, the presence of necrosis and cellular pleomorphism. These findings however, do not necessarily result in a clinically malignant behaviour. A single nasal Solitary fibrous tumour case described in the literature manifested malignancy, but was resected with no relapse.\(^{(7)}\)

CONCLUSION: Solitary fibrous tumours although uncommon, should be remembered in the differential diagnosis of nasal cavity neoplasms. The definitive diagnosis is established by immunohistochemical tests and histopathology. These reliable and accurate diagnostic criteria for Solitary fibrous tumour are necessary to avoid overdiagnosis or confusion with more aggressive neoplasms in these locations.

REFERENCES:

Fig.1: C.T scan of paranasal sinus (Axial view) showed a smooth soft tissue mass in the posterior ethmoidal sinus.
Fig. 2: C.T scan of paranasal sinus (Coronal view) showed a smooth soft tissue mass in the anterior ethmoidal sinus.

Fig. 3: Section shows spindle cells arranged in sheets and bundles with areas in between showing sclerosing band of collagen and thin vascular spaces. Cells are with scanty eosinophilic cytoplasm and elongated dark nuclei.