CASE REPORT
Volume-6 | Issue-11 | November-2017 | ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

SOLITARY FIBROUS TUMOUR OF THE NASAL CAVITY, A RARE CASE REPORT.

ENT

Dr. Manaswini Mallik MS (ENT), Department of ENT and Head Neck Surgery, All India Institute of Medical Sciences, Bhubaneswar, Odisha, India

Dr. Pradeep Pradhan MS (ENT), Department of ENT and Head Neck Surgery, All India Institute of Medical Sciences, Bhubaneswar, Odisha, India

Dr. Swagatika Samal* MD (Pathology), Department of Pathology and Lab Medicine, All India Institute of Medical Sciences, Bhubaneswar, Odisha, India *Corresponding Author

Dr. Pradipta Kumar Parida MS (ENT), Department of ENT and Head Neck Surgery, All India Institute of Medical Sciences, Bhubaneswar, Odisha, India

ABSTRACT

Solitary fibrous tumour (SFT) is a rare benign tumour in the commonly located in the pleural cavity, but extrapleural sites have been described in the literature. Again, the involvement of nose and paranasal sinuses is very rare. We report a SFT in a 43 year old female patient presented with unilateral nasal obstruction, finally diagnosed as SFT in the histopathology.

KEYWORDS

Introduction:

Solitary fibrous tumour (SFT) a rare spindle cell tumour of mesenchymal origin, is sub classified under existing mesothelial tumors. Origin of SFT is debatable, as initially named as local mesothelioma, and thought to be arising from the mesothelium; however, later studies demonstrated their origin from submucosal fibroblast-like cells. SFT was first reported in pleura by Klemper and Rabin in 1931. (1). However various extrapleural sites have been reported in the literature. Head and neck constitute only 12to 15% of all cases. In the nose and paranasal sinus, it is very rarely encountered. Definitive diagnosis is established by histopathological and immunohistochemistry study. Excision of the mass with adequate normal margin is the gold standard of treatment offered to patients with SFT. Although most cases of SFT are benign, few cases can be locally invasive and have potential to be malignant. In view of slow-growing nature of the disease, long-term follow up is needed to rule out any local recurrence.

Case report:

A 43 year old female presented with progressive nasal blockage with mucopurulent nasal discharge from the right side nasal cavity for one year. There was no history of epistaxis, epiphora, visual disturbances, headache or facial pain. Rigid nasal endoscopy revealed a firm, polypoidal mass filling the right nasal cavity. There was no cervical lymphadenopathy on clinical examination. Noncontrast computed tomography (CT) showed a homogenous isointense lesion in right side nasal cavity without any involvement of the paranasal sinuses as shown in Fig1.

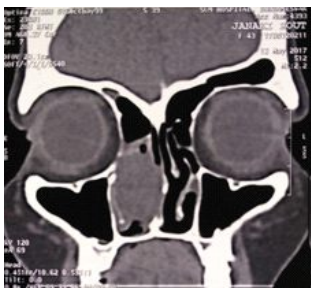

Fig 1. Computed tomography (coronal section) shows homogenous isointense polypoidal lesion in the right nasal cavity.

Routine investigations were found to be normal. Histopathological examination of the endoscopic punch biopsy revealed, spindle-shaped cells arranged in a patternless array with stromal hyalinization and thin walled, prominent branching vasculature. There was no mitosis or necrosis detected in the microscopic examination (Fig 2).

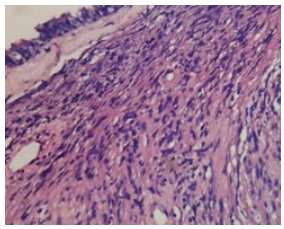

Fig 2. Histopathology shows patternless array of spindle shaped cells set in a collagenous stroma (H&E stain at 400x magnification).

On immunohistochemistry, the tumour cells were positive for STAT6 and beta-catenin while negative for CD 34 was found to be confirmed to be solitary fibrous tumour (Fig 3 and Fig 4).

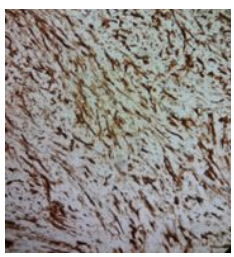

Fig 3. On immunohistochemistry, the neoplastic spindle cells are diffusely immuno positive for STAT 6.

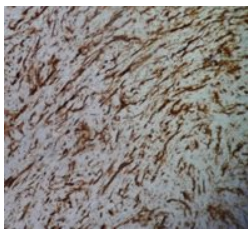

Fig 4. On immunohistochemistry, the neoplastic spindle cells are diffusely immuno positive and beta Catenin.(400x magnification)

The patient had undergone endoscopic excision of the mass under

general anaesthesia and complete excision of the mass has been achieved. The final excisional biopsy has also revealed the similar feature to that of preoperative findings without the involvement of the margin. The patient is on regular follow up for the past 6 months and is found of asymptomatic, both clinically and radiologically without any recurrence of the disease.

Discussion:

Though solitary fibrous tumour exclusively found in pleura, extrapleural sites may be affected by the tumour (0.6% of all soft tissue tumours) [2]. Various sites that may be involved in head and neck region are meninges, oral cavity, orbit, upper respiratory tract, nasal cavity, paranasal sinuses, salivary glands, thyroid, parapharyngeal space and tongue[3]. Nasal and paranasal sinuses are very rare sites of origin of SFT. 29 cases of sinonasal SFT have been reported till now in the literature [4]. SFT generally occur in 3rd to 4th decade of life, although age group between 9 to 86 years have been reported in the literature. The presenting symptoms depend upon the site of involvement. Clinically present with unilateral nasal mass, nasal obstruction, rhinorrhoea, headache, epistaxis, facial pain [5]. Larger tumours can extend intracranial by eroding cribriform plate, can goes to orbit causing exophthalmos, and can involve infratemporal fossa. Two paraneoplastic syndromes, Doege-Potter syndrome and hypertrophic osteoarthropathy can be associated with the solitary fibrous tumour. Clinically mass usually seen to be reddish, fibrous, vascular mass. In the literature, reported Size of the tumour usually measures 2 to 8 cm in major axis [5, 6].

Noncontrast CT shows homogenous isoattenuation compared with gray matter, and usually, shows enhancement after administration of contrast [6]. Occasionally they show internal calcifications. On T1 weighed images these lesions appear homogenously isointense to gray matter, compared to T2 weighed images where they appear heterogeneously isointense or hypointense. Differential diagnosis of the sinonasal solitary fibrous tumour is, nasopharyngeal angiofibroma, nasopharyngeal carcinoma, hemangiopericytoma, benign and malignant fibrous histiocytoma and fibrosarcoma [7].The definitive diagnosis of SFT is established by histopathological features and specific immunohistochemical markers. Typical histological features of SFT will show spindle-shaped cells arranged in a patternless array with stromal hyalinization and thin walled, prominent branching vasculature [8]. Though, immunohistochemistry positive for CD 34 strongly favours the diagnosis of SFT, it can be negative in 5 to 10 % of SFT [9]. Negative CD 34 usually seen in high-grade foci, recurrent tumour, or in cases of malignant transformation. Recently immunohistochemistry for STAT6 positive is found to be highly specific and sensitive for SFT. In our case, immunohistochemistry shows CD 34 negative and STAT6 positive. Presence of necrosis, 4 or more mitotic figures per 10 high power microscopic fields, increased cellularity and cellular polymorphism indicates the malignancy. Tumour size more than 10 cm, the presence of haemorrhage, infiltrative margins, and presence of anaplastic foci also favors the diagnosis towards malignancy. Though CD 34 is negative, the other features of malignancy were absent in our patient. The preferred treatment approach for sinonasal STF is endoscopic excision of mass. Other approaches like lateral rhinotomy, medial maxillectomy, anterior ethmoidectomy, sphenoidectomy has also been described. Nasal and extrapleural SFT are usually benign, whereas 23% pleural SFT have the tendency for malignancy [10]. 6 % of cases of SFT can show local recurrence after excision where as in malignancy recurrence rate can be up to 23%. Prognosis depends upon the completeness of excision of the tumour. Radiotherapy is the preferred choice of treatment in cases of malignancy, with positive margin or with metastasis. In our patient, endoscopic excision was preferred as the disease was only limited to right side nasal cavity without any destruction of bone. Long-term follow up is necessary, as the tumour has a tendency to recur.

Conclusion: Involvement of nose and paranasal sinuses by solitary fibrous tumour is very rare. Although, diagnosed by histopathological and immunohistochemistry study, a complete clinico-radilogical correlation is vital to rule out the malignancies and for the final diagnosis.

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