CASE REPORT

FIBROSARCOMA OF MAXILLARY SINUS. A RARE CASE REPORT

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ABSTRACT

Fibrosarcoma is rare malignant neoplasm, which most commonly involves the extremities. Primary fibrosarcomas of the head and neck are very rare. Only one percent of fibrosarcomas arise in these regions. The paranasal sinus is one of the rarest locations for fibrosarcoma, with only a few cases having been reported in the literature since the year 2000. Although basic information related to fibrosarcoma has been reported, there are only few previous studies which describe the imaging characteristics of paranasal sinus fibrosarcoma. So it is very challenging for a radiologist to correctly diagnose fibrosarcomas pre-operatively. Therefore we present a case of 65 years old female patient with complaints of right sided facial swelling, pain and ipsilateral nasal blockage. She underwent CT scan which showed polypoidal mucosal thickening involving right maxillary sinus containing few calcific foci within it and showing post contrast enhancement suggestive of soft tissue mass. Biopsy revealed features of low grade fibrosarcoma a very rare entity found in this region.

Key words: Fibrosarcomas, Paranasal Sinus Neoplasm, Maxillary Sinus, Maxillary Sinus Neoplasm

Introduction

Fibrosarcoma is an uncommon malignancy, with the extremities being the most common site of involvement.[1-6] Primary fibrosarcomas of the head and neck are very rare. The paranasal sinus is one of the rarest locations for fibrosarcoma, with only a few cases reported in the literature since the year 2000,[2] and less than 50 cases of fibrosarcomas arising in the maxillary sinuses have been reported up till now.[4] It is more common among males and in the 3rd and 4th decades of life.[2] Previous trauma, nasal polyposis, and a history of radiation therapy have been proposed as primary risk factor, but the etiology still remains uncertain.[3] The tissue origin of paranasal sinus fibrosarcoma appears to be the periosteum rather than mucosal connective tissue. Here we present a case of 65 years old female patient who came to ENT OPD with complaints of right sided facial swelling, pain and ipsilateral nasal blockage. She underwent CT scan which showed polypoidal mucosal thickening involving right maxillary sinus containing few calcific foci within it, lesion is showing post contrast enhancement suggestive of soft tissue mass. Patient underwent biopsy which showed features of low grade fibrosarcoma.

Case Report

A 60 year old female patient came to ENT OPD with complaints of right sided facial swelling, pain and ipsilateral nasal blockage. On physical examination there was generalized right facial fullness and numbness, however with intact right facial nerve. Inspection of the right nasal cavity demonstrated a polypoid mass. She underwent CT scan paranasal sinuses which showed polypoidal mucosal thickening involving right maxillary sinus causing its complete opacification. It contains few tiny calcific foci and showing post contrast enhancement suggestive of soft tissue polyp with suspicion of malignancy. It was

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also bulging into right nasal cavity. This was resulting in pressure erosion of anterior, superior and lateral wall of maxillary sinus.There is no evidence of intra-cranial or intra-orbital extension. Patient underwent biopsy which showed fragments of spindle cell neoplasm without capsule. Some of the fragments were partly covered by respiratory epithelium. There was moderate degree of pleomorphism. No definite mitosis seen. These appearances were suggestive of low grade fibrosarcoma which is very rare in this location.

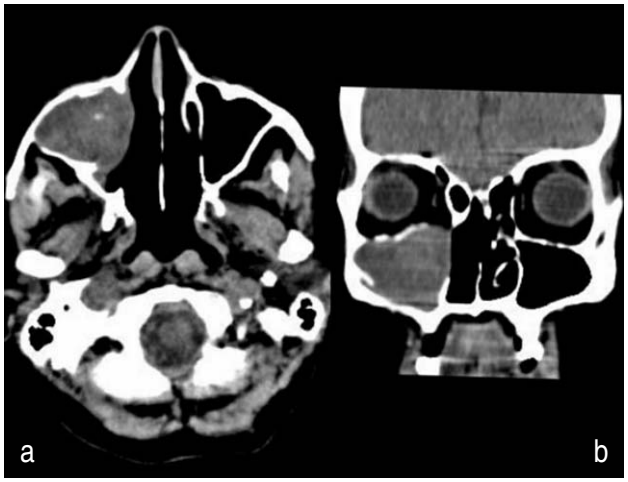

Figure 1 (A and B): Axial and coronal unenhanced CT scan images showing heterogenous lobulated soft tissue density lesion involving right maxillary sinus also bulging into right nasal cavity with few patchy areas of low density and foci of calcification.

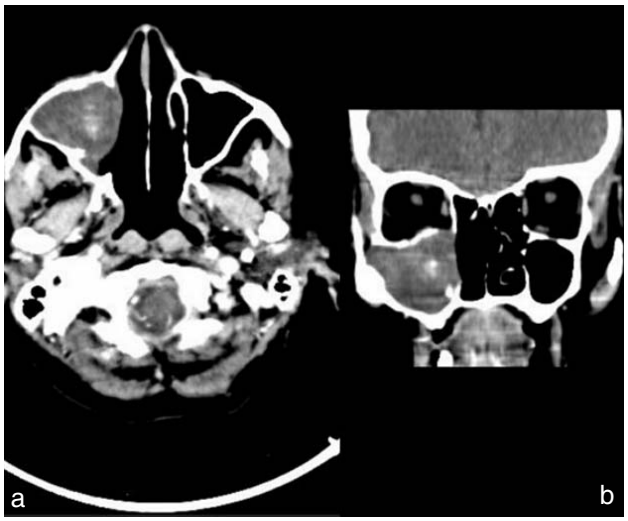

Figure 2 (A and B): Axial and coronal post contrast CT scan images showing patchy post contrast enhancement of right maxillary soft tissue lesion.

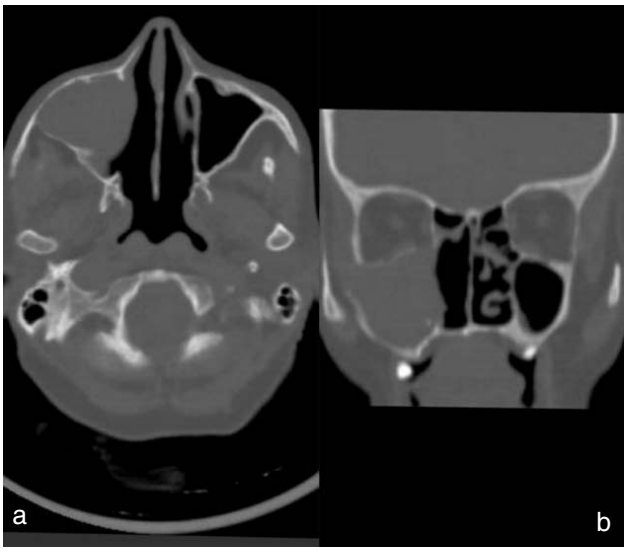

Figure 3 (A and B): Axial and coronal CT scan bone window images showing pressure erosion of walls of right maxillary sinus.

Discussion

Fibrosarcoma is a malignant tumor of fibrocytes which is seen very rarely in the region of nasal cavity and paranasal sinuses. Previous trauma, nasal polyposis, and a history of radiation therapy have all been implicated as frequent risk factors, but the definite etiology still remains unclear.[3] The clinical symptoms of paranasal sinus fibrosarcoma are very non-specific and may include pain in the cheek as well as in head, nasal obstruction, epistaxis, hypoesthesia of the facial region, a mass in the maxillary area, and chronic sinusitis.[1,3,5] Ocular, auditory, facial and oral symptoms can also rarely occur.

Radiography show bony erosion as well as soft tissue opacification of the sino-nasal region.[3] On CT Paranasal sinus fibrosarcomas appear as heterogeneous lobulated, soft-tissue density masses with patchy areas of low density, foci of calcification and may show post contrast enhancement. On MR imaging fibrosarcomas tended to be iso to hypointense on T1 weighted images, and may appear heterogeneous mild hypointense on T2 weighted images, suggestive of high cellularity, high nuclear-to-cytoplasm ratio and the presence of abundant fibrous tissues. Bone destruction is a common feature of sino-nasal fibrosarcoma.[1]

On histopathology fibrosarcomas are encapsulated, spindle cell tumors. Low-grade sino-nasal fibrosar-

coma is slow growing, resulting in compression of the adjacent bony structures, while poorly differentiated paranasal sinus fibrosarcoma presents an invasive growth pattern because of the greater degree of pleomorphism and nuclear atypia followed by bone erosion.[1] Pathological diagnosis of paranasal sinus fibrosarcoma depends on the presence of a characteristic "herringbone" pattern consisting of elongated spindle-shaped cells and positive markers for vimentin or CD34.

Sinonasal fibrosarcoma is a highly malignant neoplasm with rare distant metastasis and frequent local recurrence. Therefore, local excision with large surgical borders has importance in treatment.[2] It is resistant to chemotherapy. Surgery is the preferred treatment approach. Large local excision is the most commonly used approach. Adjuvant radio-therapy may be applied in the presence of surgical border involvement or when the tumor cannot be fully removed.[2] Death usually results from intracranial extension of a persistent or recurrent tumor.[3]

On the basis of imaging alone, the differential diagnosis of an aggressive mass arising in a paranasal sinus may include squamous cell carcinoma, minor salivary gland tumors, primary bone tumors, metastatic deposits, lymphoma, and plasmacytoma. Occasionally, benign diseases such as aggressive polyposis, inverting papillomas, and mucoceles may present with bone erosion which is an important feature of fibrosarcomas.[3]

Conclusion

Being a challenging entity for a radiologist to diagnose sino-nasal fibrosarcomas on imaging alone however it should be considered in the differential diagnosis when a sino-nasal neoplasm appears as a well or ill-defined unilateral mass with foci of calcification and enhancement on CT and shows characteristic mild hypointensity on T2 weighted MR images with heterogeneous delayed post contrast-enhancement pattern along with expansile or osteolytic bony destruction.

Conflict of Interest: None to declare

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