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Nasal Cavity Malignant Solitary Fibrous Tumor: A Case Report

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ABSTRACT

A solitary fibrous tumor (SFT) is a soft tissue sarcoma that arises from soft tissue cells and can arise anywhere in the body. Their incidence is less than 2% of all soft tissue masses. They are generally found in the pleural cavity, but sometimes extra-thoracic sites are also involved. Malignant SFTs of the nasal cavity are very rare. The paper reports a case of a 58-year-old male patient with complaints of left nasal blockage and mass. The patient underwent Functional Endoscopic Sinus Surgery (FESS), and the excised mass was sent for histopathological examination, which showed features consistent with malignant solitary fibrous tumor with CD34, CD99, and BCL2 positive. The patient was started on Adjuvant chemotherapy, has received 2 cycles of chemotherapy, and is currently under treatment.

1. Introduction

A solitary fibrous tumor (SFT), also known as benign fibrous mesothelioma or sub mesothelial fibroma,[1] is a rare soft tissue tumor of mesenchymal origin. The true nature of these tumors is controversial but is now believed to be mesenchymal in origin, as evident by their fibroblastic differentiation immunohistochemically, histopathologically, and structurally.[2] This explains why SFTs are found in various extrapleural sites like orbit, meninges, peritoneum, pelvis, liver, adrenal gland, and urogenital tract.[3-5] SFTs are also found in the head and neck region, but their occurrence in the nose and nasal cavity is extremely rare. They are generally asymptomatic initially, with symptoms arising as the mass grows in size. When a malignant SFT is found, the primary treatment of choice is adequate resection with negative margins for successful treatment.

2. Case presentation

A 58-year-old male patient with a history of hypertension attended the Department of Otorhinolaryngology, Regional Institute of Medical Sciences (RIMS) with a complaint of left nasal blockage for the last year and left nasal mass for the last nine months. The nasal blockage was limited to the left side with a history of non-foul smelling, watery nasal discharge. The nasal mass was initially small and gradually increased over time to become visible nine months back. There was no history of epistaxis, headache, sinusitis, watering of eyes, diplopia, epiphora, or change in visual acuity. There is no family history of cancer or similar illness in the past. The patient presented late due to the fear of getting infected with the COVID-19 virus and received only symptomatic treatment. The patient gives a history of occasional alcohol consumption (2-3 peg/week) for the past 25-30 years, with betel nut chewing for more than 30 years. There is a history of pulmonary tuberculosis 20 years back, which was cured after completing the course of anti-tuberculosis treatment and is Acid Fast Bacillus (AFB) negative at present.

On physical examination, a polypoid mass (1.5 × 2 cm in size) was seen from the left lateral nose wall with deviated nasal septum towards the right side. A thorough cranial nerve examination was done and was found to be unremarkable. Contrast-Enhanced Computed Tomography (CECT) scan of the nasal cavity and Paranasal Sinus (PNS) showed a large expansile mass (1.8 × 2.5 × 2.0 cm) confined to the left nasal cavity with a deviation of the nasal septum towards the right side, with features of sinusitis (Fig. 1). There was no intracranial extension or bony erosion. The patient underwent Functional Endoscopic Sinus Surgery (FESS) and Septoplasty, and the excised specimen was sent for histopathological examination (HPE).

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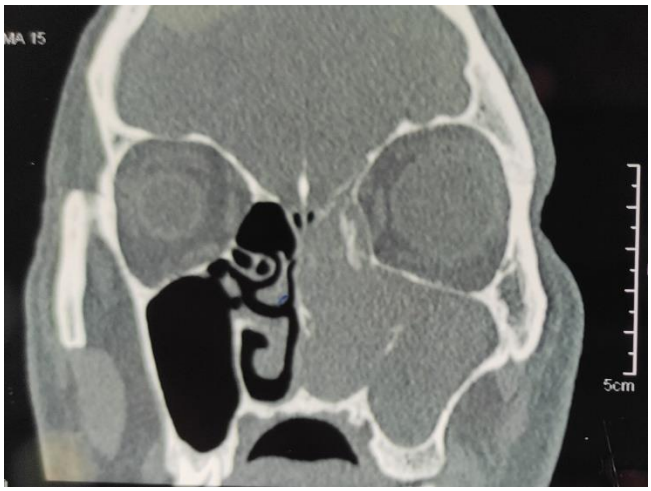

Fig. 1. CECT scan of nasal cavity and PNS (coronal view) showing a large expansile mass in the left nasal cavity with pansinusitis.

HPE of the excised specimen showed features suggestive of malignant solitary fibrous tumor (malignant hemangiopericytoma like a tumor) with no evidence of necrosis or dystrophic calcifications, with an abnormally high mitotic activity [(8-10/10 High Power Field (HPFs)] and negative surgical margins (Fig. 2). Immunohistochemistry (IHC) studies were strongly positive for Cluster of differentiation 34 (CD34), CD99, and B-cell lymphoma 2 (BCL-2) (Figs. 3 and 4); and negative for CD31 and Cytokeratin. After confirming the diagnosis, the patient was transferred to the Department of Radiation Oncology, RIMS. A thorough physical examination and metastatic workup were done. On examination, patients were in very good general condition, with body surface area being 1.6 m^2 and Karnofsky Performance Score (KPS) being 90%. No peripheral lymph nodes were clinically palpable.

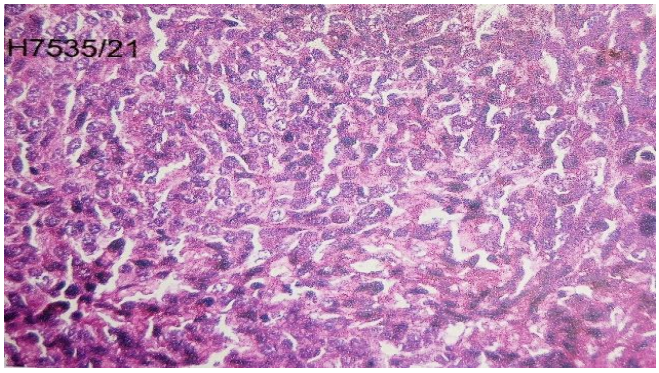

Fig. 2. HPE of the excised nasal cavity specimen showing features suggestive of a solitary fibrous tumor.

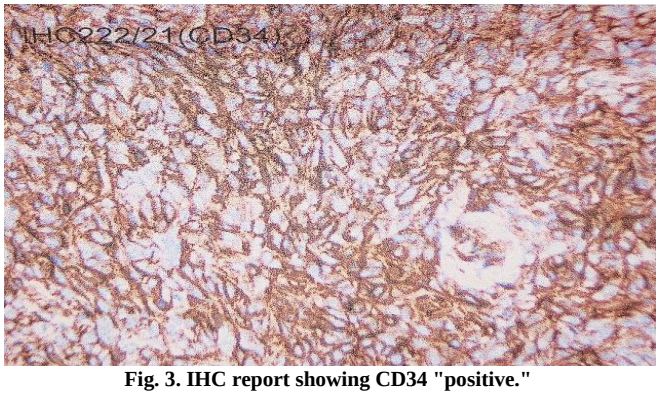

Fig. 3. IHC report showing CD34 "positive."

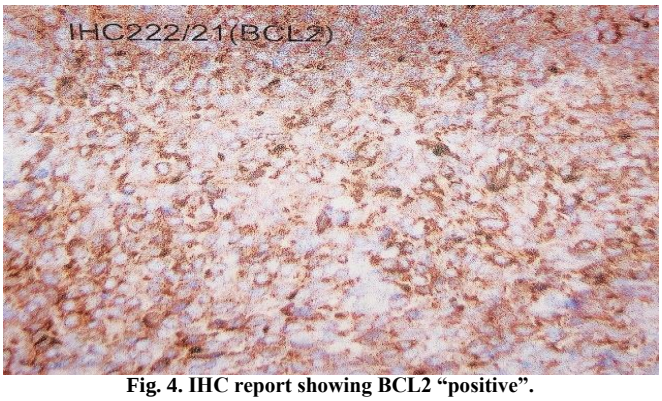

Fig. 4. IHC report showing BCL2 "positive".

Routine baseline investigations (Complete Blood Count, biochemistry and Electrocardiogram) were done and were found to be uneventful and metastatic workup (Chest X-ray and Ultrasound of the whole abdomen) found no sites of metastasis or any organ involvement. Postoperative CECT scan of nasal cavity showed no residual disease with features suggestive of the mild deviated nasal septum on the right side with peri sinusitis (Fig. 5). The patient was started on single-agent adjuvant chemotherapy with the injection of Pegylated Liposomal Doxorubicin (80 mg), four-weekly cycles. The patient has received two cycles of chemotherapy till now and is currently under treatment.

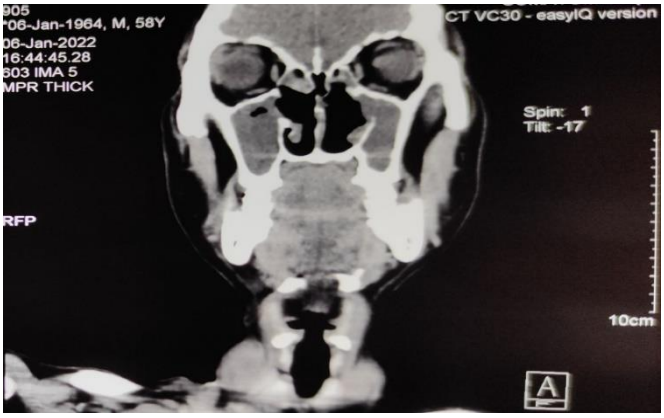

Fig. 5. Post-op CECT scan of the nasal cavity (coronal view) showing no residual disease and deviated nasal septum.

3. Discussion

Solitary Fibrous Tumors (SFTs) are rare tumors of mesothelial origin that occur mainly in the third and fourth decade of life, with a range of onset ranging from 9 to 86 years.[6] They were believed to be exclusively located in the thoracic cavity as a pleural tumor,[7] but found in various extrathoracic sites, including the head and neck region. The mesenchymal origin of the tumor is supported by the growth of the tumor in various extra serosal regions, with extrathoracic SFTs having histological, immunohistochemical, and structural features similar to those tumors that arise from pleura-based intrathoracic region.[8] Extrathoracic SFTs are generally benign, with approximately 10-15% of extrathoracic SFTs being recurrent or metastatic. Only a small percentage of histologically benign tumors show clinically malignant behavior.[5] Local recurrence or metastasis are rare, and it depends on the size of the tumor, infiltration into the surrounding tissues, surgical margins, necrosis, and mitotic activity. Nasal SFTs initially present as an asymptomatic, well-circumscribed, painless, slow-growing soft tissue mass. After that, as the tumor increases in size, various other symptoms develop, such as nasal obstruction, sinusitis, headache, epistaxis, rhinorrhea, hyposmia,

exophthalmos, facial pain, or visual disturbance.[9, 10] The diagnosis of SFTs depends on a detailed history, thorough clinical examination, CT (Computed Tomography)/MRI (Magnetic Resonance Imaging) scan, and histopathological examination with immunohistochemistry. These tumors are usually red, fibrous, and well encapsulated. Differential diagnosis of SFTs is very difficult as these tumors show histological features similar to those found in other soft tissue neoplasms such as hemangiopericytoma, nasopharyngeal angiofibroma, fibrosarcoma, fibrous histiocytoma, hemangioma, inverted papilloma, nasopharyngeal carcinoma, and schwannoma.[11] Macroscopically, SFTs are polypoidal, well-circumscribed, sessile or pedunculated, tan rubbery mass.[12] Microscopically, SFTs are low-grade neoplasms with a characteristic "patternless" pattern and variable cellularity, with a random arrangement of spindle cells, hypo or hypercellular sclerotic foci, stromal hyalinization with hemangiopericytoma pattern.[8] Immunohistochemistry is important for the diagnosis of SFTs. They are strongly positive for CD34, CD99, BCL2, and Vimentin. They are negative for CD31, Cytokeratin, Epithelial Membrane Antigen (EMA), S-100 protein, Glial fibrillary acidic protein, Desmin, and Carcinoembryonic Antigen (CEA).[9, 13] Study conducted by Gold et al. showed tumor features associated with malignant behavior, including recurrent tumor, macro- or microscopically positive resection margin, increased cellularity and nuclear pleomorphism, tumor size more than 10cm, more than four mitoses/10 HPFs, and presence of malignant components.[14] There is no globally accepted treatment strategy, due to which a multidisciplinary team consisting of surgeons, medical oncologists, and radiation oncologists are involved in the management of SFTs. Complete surgical resection is the treatment of choice for a nasal cavity or paranasal sinus SFTs, with the endoscopic method being the preferred surgical approach.[9] Resectability is an important prognostic factor. Thus, an en bloc surgical excision is preferred to treat SFTs of the nasal cavity and paranasal sinuses effectively.[6, 15, 16] The use of radiation therapy in the treatment of SFTs is unclear, and its use is decided on a case-by-case basis as a part of a multidisciplinary plan. Due to the low incidence of SFTs, choosing the most effective chemotherapy for advanced SFT is very difficult.

4. Conclusions

A solitary Fibrous Tumor is a slow-growing mesenchymal tumor. SFTs involving the nose and paranasal sinuses are rare, with extremely rare malignant SFTs. Proper histopathological examination (HPE) and immunohistochemistry (IHC) analysis is required for the correct diagnosis of SFTs. Our patient was diagnosed as a malignant solitary fibrous tumor of the nasal cavity and paranasal sinuses based on HPE and IHC of the excised specimen from the nasal cavity and is currently undergoing treatment with a single agent chemotherapy regular follow-up.

Conflict of Interest

The authors declared that there is no conflict of interest.

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