Case Report / Olgu Sunumu

Sinonasal inflammatory myofibroblastic pseudotumor (plasma cell granuloma)

Sinonazal enflamatuvar miyofibroblastik psödotümör (plazma hücreli granülom)

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ABSTRACT

Inflammatory myofibroblastic pseudotumor (plasma cell granuloma) is a soft tissue lesion consisting of myofibroblasts, mature lymphocytes, histiocytes, plasma cells, eosinophils, and extracellular collagen. Various sites in the body may harbor these lesions. Lungs, omentum, intestines, mesentery, and urinary system are the most susceptible areas. It is usually seen in children and young adults. The lesion is rarely detected in the head and neck region. The orbit and the upper respiratory system are the most common localizations in the head and neck region. Sinonasal tract is a rare site of involvement. The differential diagnosis includes squamous cell carcinoma (spindle cell variant), inflammatory fibrosarcoma, leiomyosarcoma, schwannoma, and nonspecific inflammation. Our patient who had a sinonasal mass showed a benign tumor consisting of spindle tumor cells and inflammatory cells histopathologically. This case was presented due to its rare existence to this site.

Keywords: Inflammatory myofibroblastic pseudotumor; plasma cell granuloma; sinonasal.

ÖZ

Enflamatuvar miyofibroblastik psödotümör (plazma hücreli granülom) miyofibroblastlar, olgun lenfositler, histiositler, plazma hücreleri, eozinofiller ve ekstraselüler kollajenden oluşan bir yumuşak doku lezyonudur. Bu lezyonlar vücuttaki çeşitli bölgelerde bulunabilir. Akciğerler, omentum, bağırsaklar, mezenter ve üriner sistem riske en açık alanlardır. Genellikle çocuklar ve genç erişkinlerde görülür. Lezyon baş ve boyun bölgesinde nadiren tespit edilir. Baş ve boyun bölgesinde orbit ve üst solunum sistemi en yaygın yerleşim yerleridir. Sinonazal yol nadir bir tutulum bölgesidir. Ayırıcı tanı skuamöz hücreli karsinom (iğsi hücreli varyant), enflamatuvar fibrosarkom, leiomiyosarkom, schwannom ve nonspesifik enflamasyonu içerir. Sinonazal kitlesi olan hastamızda histopatolojik olarak iğsi tümör hücreleri ve enflamatuvar hücrelerden oluşan iyi huylu bir tümör görüldü. Bu olgu bu alandaki nadir oluşumu nedeniyle sunuldu.

Anahtar Sözcükler: Enflamatuvar miyofibroblastik psödotümör; plazma hücreli granülom; sinonazal.

Inflammatory myofibroblastic pseudotumor (IMPT) of the head and neck is a rare entity. It is considered a stromal tumor with undetermined aggressiveness related to inflammatory fibrosarcoma.[1] Several different names such as plasma cell granuloma, plasma cell pseudotumor, inflammatory myofibroblastic tumor, inflammatory fibrosarcoma and most commonly

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inflammatory pseudotumor have been used to describe this lesion. Although the etiology and pathogenesis of these lesions are still not clear, an exaggerated inflammatory reaction to tissue injury of unknown causes is held responsible.[2] Inflammatory myofibroblastic pseudotumor can mimic expansile and invasive malignant tumors, both clinically and radiologically. A case of IMPT with bilateral sinonasal and paranasal localization was presented to our department. Wide surgical excision was performed under endoscopic guidance. In this report, we present a rare case of sinonasal IMPT and discuss its diagnosis and treatment.

CASE REPORT

A 53-year-old man was admitted to the Mersin University Medical School with symptoms of nasal obstruction, pain and swelling of the face over a duration of six months. Physical examination revealed bilateral nasal polyposis mimicking inverted papilloma. He had no cervical lymphadenopathy and the remaining physical examination was unremarkable. Computed tomography (CT) revealed a soft tissue mass involving bilateral maxillary and ethmoidal sinuses. The mass was expanding the lumen and the ostium of the right maxillary sinus and was extending into the nasal cavities, especially on the right side. Bone remodeling at the medial wall of the right maxillary sinus was also noted (Figure 1).

Endoscopic examination performed before surgery revealed a polypoid mass completely filling both nasal cavities. All surrounding soft tissues were excised under endoscopic guidance (Figure 2). The maxillary sinuses were opened bilaterally and the polypoid soft tissues were excised. Anterior and posterior ethmoidectomy was performed and the ostia of sphenoid and frontal sinuses were explored. These sinuses were free of soft tissue lesions.

The resection specimen consisted of a reddish mass due to extensive hemorrhage. Grossly, the masses were tan-white and glistening, measuring 7x6x3 and 6x5x3 cm diameters for the right and left maxillary sinuses respectively. Pathological examination showed a benign tumor consisting of a proliferation of spindle tumor cells, which had accompanying inflammatory cells--predominantly plasma cells, histiocytes, lymphocytes and multinuclear giant cells (Figure 3a). Small lymphoid follicles in the fibrous tissue were also noted. Necrosis and mitoses were not detected. No fungal organisms were seen with both periodic acid-schiff (PAS)

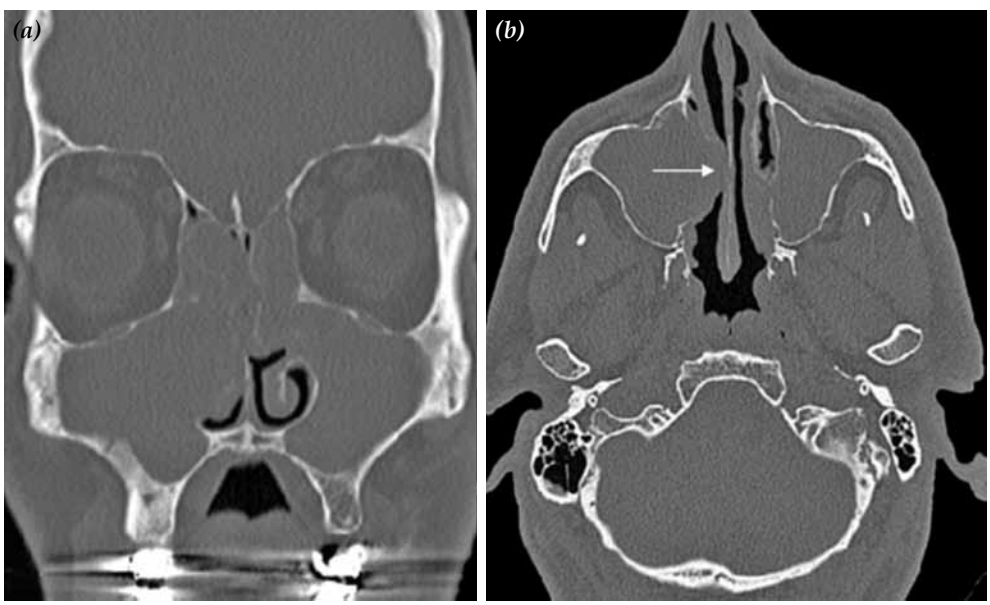

Figure 1. *Coronal computed tomography scan* ***(a)*** *shows a large soft tissue mass occupying both right and left maxillary and ethmoidal sinuses extending into both nasal cavities. Axial computed tomography scan* ***(b)*** *better demonstrates bone remodeling (arrow) at the medial wall of the right maxillary sinus.*

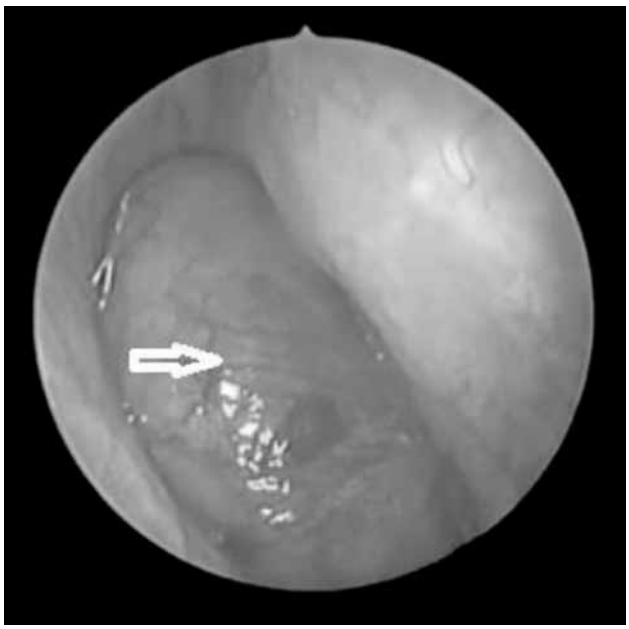

Figure 2. *Polypoid mass (arrow) seen in the right nasal cavity during nasal endoscopic examination.*

and silver staining. Immunohistochemical studies showed that the spindle tumor cells were positive for vimentin and smooth muscle actin (SMA) (Figure 3b) whereas they were negative for desmin, S-100, cytokeratin (CK), and CD34. The plasma cells showed positive staining with CD138. Monoclonality was not detected with immunohistochemistry of the plasma cells; which positively stained Lambda and Kappa (Figure 3c, d). The lymphocytes were positive for leukocyte common antigen (LCA) and had polyclonal staining with both CD3 and CD20. Histiocytes were positive for CD68 and alpha1-antitrypsin (α1-AT). Based on these findings, a final diagnosis of inflammatory myofibroblastic pseudotumor was confirmed.

Histologically, the surgical resection margin was free of tumor. The recurrent control biopsy taken 4-5 months after surgery showed only chronic inflammation on histopathological examination. The postoperative period was uneventful and follow-up of the patient was free of tumor for one year.

DISCUSSION

Inflammatory myofibroblastic pseudotumor is a benign, tumor like mass characterized by proliferating fibrous tissue infiltrated by inflammatory cells.[3] This condition is known to

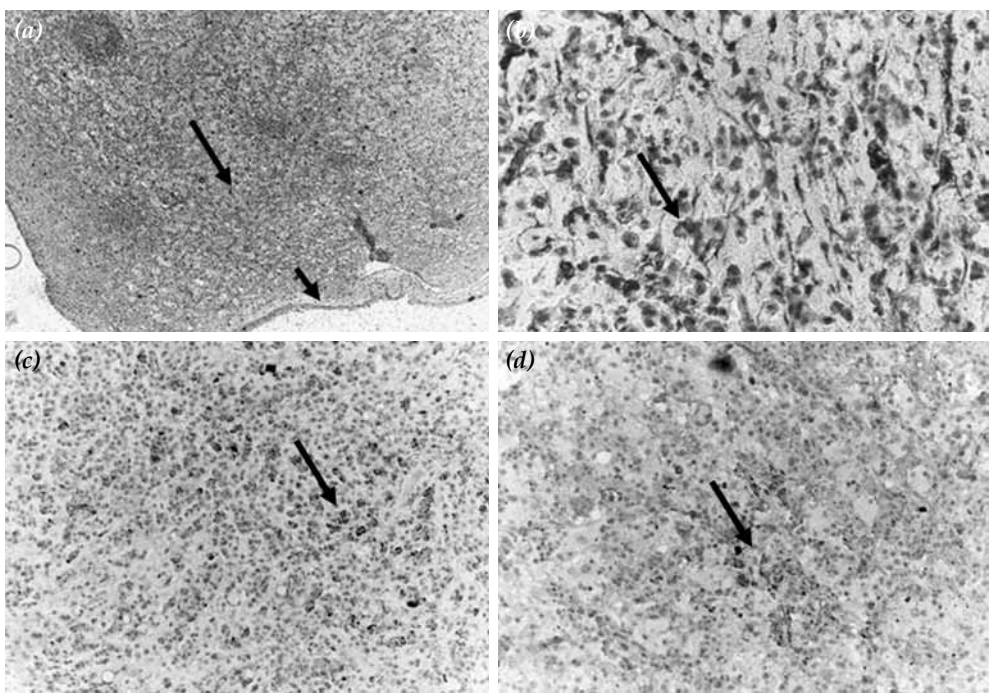

Figure 3. ***(a)*** *Tumor including inflammatory cells (long arrow) and spindle cells covered by respiratory epithelium (short arrow) (H-E x 40),* ***(b)*** *The spindle tumor cells (arrow) were positive for antibody to smooth muscle actin (SMA x 400),* ***(c)*** *Polyclonal staining of plasma cells (arrow) for Lambda (Lambda x 200),* ***(d)*** *Polyclonal staining of plasma cells (arrow) for Kappa (Kappa x 200).*

occur in several organs including lymph nodes, spleen, brain, spinal cord, larynx, thyroid gland, breast, pancreas, gastrointestinal (GI) tract and bladder, but occurs most frequently in the lungs and liver.[4] Only a few cases in the sinonasal cavities and sinuses have been reported[5] and IMPT involving the ear lobe and retroauricular region have also been reported.[6,7]

These tumors present with a variety of symptoms depending on their localization. The symptoms of IMPT in nasal cavities are non-specific. Unlike similar tumors in the visceral organs, sinonasal IMPT cause no systemic symptoms such as fever, anorexia, weight loss and malaise, but cervical and axillary lymphadenopathy have been reported.[8] They may easily be misdiagnosed in early stages. Clinical or endoscopic findings may demonstrate a lesion covered by normal mucosa, a polyp or edematous mucosa.[7] Computed tomography examination of IMPT in the sinonasal cavities shows a homogenous mass that is slightly heterogeneous if contrast material is given.[9] Magnetic resonance imaging (MRI) usually shows a mass with low to intermediate signal intensity on both T_1- and T_2-weighted imaging. Variable contrast enhancement can be seen on both CT and MRI. Bone changes mimicking malignancy such as erosion, sclerosis, remodeling and thickening can also be detected on imaging studies.[10,11] The destructive nature of sinonasal IMPT is a constant finding and is suggestive of a malignant process. One case of intracranial extension of a tumor located in the maxillary sinus was also reported.[9]

Histologically, IMPT is composed of proliferating myofibroblasts and fibroblasts, mixed with a prominent infiltrate of lymphocytes, plasma cells, and acute inflammatory cells. Three distinct histological patterns have been described-- a) myxoid/ vascular pattern, resembling inflammatory granulation tissue, b) compact spindle cell pattern with fascicular and/or storiform areas and varying cellular density, and c) hypocellular pattern, densely collagenized lesion resembling a fibrous scar tissue. These three patterns may be equally represented within the same tumor, or one or two patterns may predominate.[12] The histopathological type of our case was the first type with a myxoid-vascular pattern. Meis and Enzinger[13] reported that the recurrence rate of this type was up to 37%. In all localizations of IMPT recurrence is more common in the sinonasal cavities. Some reviews have suggested that the recurrence rate of IMPT with myofibroblastic or fibroblastic phenotype in the nasal sinus is higher than other types.[14] There is also a controversy about the nature of IMPT; a reactive process or a true neoplasm. The cases located in the sinonasal cavities have more aggressive behavior, poor prognosis and show multiple recurrences.[2] Inflammatory myofibroblastic pseudotumor can mimic a malignant tumor both radiologically and clinically. The diagnosis of inflammatory pseudotumor is one of exclusion based on clinical and imaging studies. Image findings are not characteristic. The soft tissue disease may be associated with bone erosion, remodeling, sclerosis, and thickening.[10] On magnetic resonance imaging, IMPT are usually isointense to hypointense relative to muscle on T_1-weighted images, with a relatively hypointense T_2 signal compared to most other tumors. Contrast enhancement is variable. The radiologic appearance can also be misinterpreted as a malignancy due to bone changes such as erosion, sclerosis and thickening. A preoperative diagnosis of IMPT is difficult because of the diverse clinical settings in which they arise. Diagnosis of IMPT is not possible without histopathologic examination of a tissue sample.[15]

When the clinical manifestation and imaging studies show an aggressive appearance, as it did in this case, the diagnosis can be challenging for clinicians. In general, the diagnosis of IMPT is made either from multiple incisional biopsies in the search for an underlying pathological process or from an initial excisional biopsy with a thorough investigation of morphological features. Differential diagnoses include carcinoma, fibrodysplastic lesions, lymphoma, chronic fungal infection, Wegener's granulomatosis, and extramedullary plasmacytoma.[16] And it must also be distinguished from sinusitis, granulomatous inflammation, collagen vascular diseases, sarcoidosis and neoplastic disorders.[17]

Histologic transformation to an undifferentiated sarcomatous proliferation has

not been reported in sinonasal IMPT. Unlike orbital localizations, sinonasal IMPT never transform into lymphoma or develop into systemic lymphoma.[18]

Despite initial aggressive clinical and radiologic presentations, sinonasal IMPT has a good prognosis. Surgery, corticosteroids, radiotherapy and chemotherapy have been used in the treatment of these tumors.[19]

Inflammatory pseudotumor is a rare disease in the sinonasal area. Because of its aggressive appearance, differential diagnosis with malignancy is warranted. Multiple biopsies are needed to get a definite diagnosis. It is vital that rhinologists be familiar with the clinical presentation due to similarities with malignant tumors.[20] Its correct recognition by the pathologist is vital to avoid unnecessary treatment such as extensive surgical intervention or radiotherapy.

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