Inflammatory myofibroblastic pseudotumor (IMT) is a rare disease entity characterized by a proliferation of fibroblasts, myofibroblasts, lymphocytes, plasma cells, and histiocytes. Although this is not a malignant tumor at cytomorphological level, its complexity in differential diagnoses and a tendency for recurrence creates challenges in clinical management, especially in the head and neck region. Reported here is a case of a recurrent IMT of the maxillary sinus with a complex clinical course. We present the data on the utility of fine needle aspiration biopsy (FNAB) along with previous surgical biopsies with immunohistochemical staining in diagnosing and monitoring IMTs, as well as a detailed discussion of the cytopathological findings and our strategies of clinical management in this patient. The findings indicate that FNAB is a valuable tool in monitoring such a patient.

INTRODUCTION

Immunohistochemistry was helpful to further clarify our patient’s tumor. The myofibroblastic cells were positive for CD68, a histiocytic marker, and actin, a myxoid marker, but negative for CAM 5.2, S-100, desmin, cytokeratin AE 1/3, CD34, ALK-1, cytokeratin 903, CD 177, and EGF. Calponin and vimentin were focally positive; both kappa and lambda light chains were positive in lymphoid population. This profile is typical for an IMT. Our case showed that 93% of IMTs are positive for calponin. 33% for desmin. 0% for S-100. 0% for CD34, and 80% for ALK-1. The negativity for ALK-1 may have clinical implications, as Coffin et al. showed that ALK negative IMTs, 46% recurred and 100% metastasized (p<0.02), while for ALK positive IMTs, 0% recurred (n=31). Diagnosis of IMT requires a histological specimen along with immunohistochemical staining. The differential diagnosis includes neoplastic growth, granulomatous diseases, vasculitis, and infections. CT scanning shows a homogenous tissue density with some enhancement. 

DIAGNOSIS

The case presented a highly recurrent and locally invasive IMT of the maxillary sinus. The tumor recurred five times, with multiple unsuccessful therapeutic intervention and severe reaction and reconstruction was performed. In the twelve-year course of her disease, the patient was treated with multiple surgeries, two courses of external beam radiation, and a one-year course of prednisone treatment. By the time of the case presentation, the patient had tumor extension into many of the anatomical structures adjacent to the maxilla, including the temporal bone, orbital contents, parotid gland, and the mandible.

IMTs can be divided into three groups based on their histologic appearance: myxoid vascular and inflammatory proliferation resembling nodular fasciitis; compact spindle cell proliferation resembling fibrous histiocyte or fibromatosis; dense collagen deposition with sparsely cellular components resembling a scar or desmoid tumor. Our patient’s IMT would be classified as the first type, as she displayed myofibroblastic proliferation and nonspecific inflammation.

The case presented showed that FNAB is a useful test for monitoring IMT recurrence.1 Our patient had two FNAs performed, and while both gave hypocellular material, there were enough cells to identify spindle cells amidst an inflammatory cellular background. Given a poor history of IMT, that cytological profile should be enough to diagnose recurrence. Furthermore, if the fine needle aspiration yields enough cells for immunohistochemical staining, its diagnostic utility is greater in biopsy, which was diagnosed as an inflammatory myofibroblastic tumor. Figure 1, H&E stain on low power with high power insert.

CASE REPORT

In December of 2006, the patient returned to the Mount Sinai hospital with right sided facial paralysis and incomplete eye closure. She underwent placement of a gold weight into the superior eyelid to achieve better closure. The patient then returned to the hospital in May of 2007 with an expanding mass in the right infraorbital fossa that was causing edema and tenderness of the right temporal region and the right eyelid. A biopsy was performed, and it showed inflammatory myofibroblastic tumor that stained negative for ALK-1, CD177, and EGF. The patient underwent a magnetic resonance imaging scan in May of 2007 that showed tumor in the infraorbital fossa and pharyngeal wall as well as in the floor and lateral wall of the orbit and extending up through the foramen ovale to involve the cavernous sinus on the right side. Based on the MRI and biopsy results, the patient was scheduled for an extensive surgery in June of 2007, which included a temporal craniectomy, radical maxillectomy, skull base resection, orbital exenteration, and a rectus abdominis free flap reconstruction. Pathology showed multiple areas of tumor recurrence. The patient’s clinical course was complicated by development of a draining abscess in the previous surgical site during September of 2007. She was treated with placement of a drain and 4 weeks IV vancomycin to cover ORSA that was cultured from the pus. Based on magnetic resonance imaging performed in February of 2008, in May of 2008, the patient underwent 2 CT scan guided core biopsies of the right carotid sheath, and the pathology showed “inflamed inflammatory cell infiltration, fibroblast/myofibroblastic proliferation, and collagen deposition, consistent with inflammatory myofibroblastic tumor.” An MRI of the neck performed on 10/18/08 showed progressive increase in size of the mass in the vicinity of the right internal carotid artery, with displacement and partial compression of the artery.

REFERENCES


