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Inflammatory pseudotumor in head and neck

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ABSTRACT

Background: Inflammatory pseudotumor (IPT) is a tumefactive lesion characterized by fibroblastic proliferations and a prominent inflammatory component. It behaves as a locally benign or aggressive lesion, clinically and radiologically mimicking a neoplastic process. Numerous entities can be diagnosed as IPT, from reactive lesions to true neoplasms. The diagnosis of IPT requires further elaboration, and IPT should be distinguished from other similar entities such as inflammatory myofibroblastic tumor and IgG4-related sclerosing disease.

Case summary: We report two cases of IPT arising from the head and neck region. One occurred at the orbit and the other at the parapharyngeal space. Histologically, they showed aggregates of myofibroblasts and inflammatory cells. Immunohistochemically, the number of IgG4-positive cells was less than 40% of the number of IgG positive cells, and the myofibroblastic cells were negative for anaplastic lymphoma kinase. The diagnosis was IPT/not otherwise specified. One patient was treated by systemic administration of corticosteroid and had good response. The other, who was treated by local administration of corticosteroid, partially responded and is currently stable with limited disease.

Discussion: IPT has been reported to occur in various anatomical sites, most commonly in the lungs. The incidence in the head and neck area is extremely rare. Treatment of IPT is controversial and may involve corticosteroids or surgical resection, or both. Other chemotherapeutic agents and radiotherapy may be considered in steroid-resistant patients. The pathological subtype, safety of resection, and safety of corticosteroid use must be included in the decision-making process for treatment.

1. Introduction

Inflammatory pseudotumor (IPT) is a heterogeneous group of lesions sharing similar histologic features of fibroblastic and myofibroblastic proliferation with inflammatory infiltrates. It often shows aggressive clinical behavior, with locally destructive or infiltrative features that mimic malignant tumors. The etiology and pathogenesis of IPT remains controversial; it contains diverse entities, from reactive lesions to true tumors. Because a diagnosis of IPT is ambiguous and can cause clinical confusion, the diagnosis of IPT should be further elaborated. Recently, inflammatory myofibroblastic tumor (IMT) has been thought to be a neoplastic counterpart of the broad category of IPT, because IMT shows cytogenetic clonal abnormality and aberrant expression of anaplastic lymphoma kinase (ALK) and its gene translocation. In addition, some cases which were previously diagnosed as IPT, have been found to be associated with IgG4-related sclerosing disease, which is a reactive lymphoproliferative-sclerosing lesion that can involve various organ systems of the body. Occasionally, it is still difficult to judge whether such ambiguous cases are reactive or neoplastic, despite various attempts to develop differentiation techniques. In such cases, the diagnosis becomes IPT/not otherwise specified (NOS), and it is necessary for pathologists and clinicians to share the recognition that a biologically-based prediction from this diagnosis is difficult. Here we report two rare cases of IPT arising from the orbit and parapharyngeal space. In each case the diagnosis was IPT/NOS. They were treated successfully with steroid therapy and maintained stable-disease status.

2. Case report

2.1. Case 1

A 74-year-old male presented with a 2-month history of right facial nerve palsy and a 1-month history of hoarseness and dysphagia. His medical history included diabetes mellitus, atrial fibrillation, and old cerebral infarction. Physical examination revealed right otitis media with effusion, complete facial nerve

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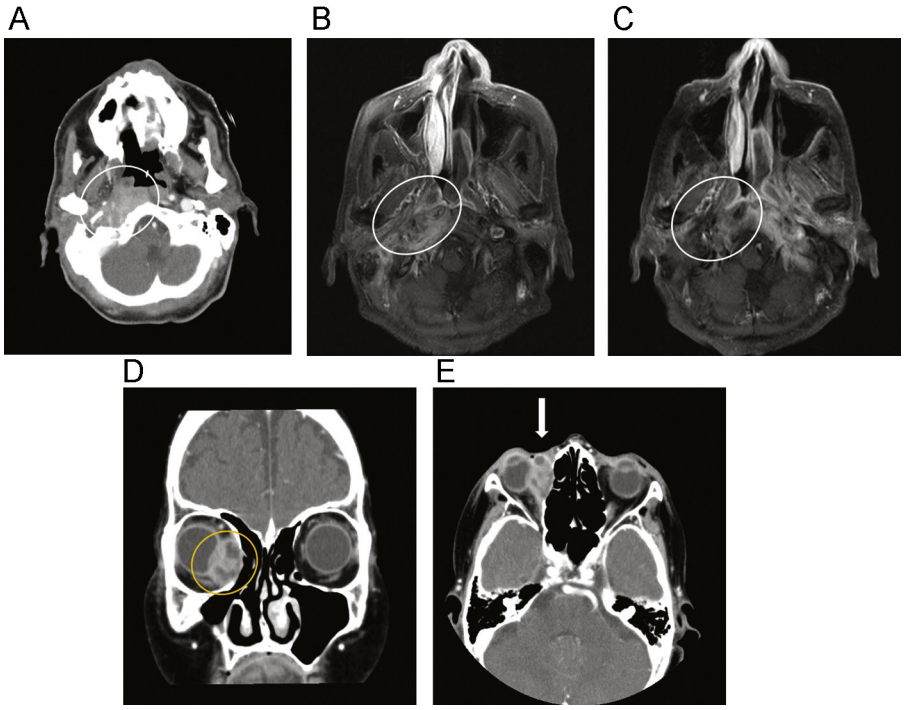

Fig. 1. CT and MRI scans. (A) CT scan of case 1 showed an enhanced lesion from the nasopharynx to the parapharyngeal space of the right side with temporal bone destruction. (B) Contrast-enhanced T1-weighted MRI showed homogeneous enhancement of the lesion. (C) Enhanced MRI 5 months after prednisolone was started. The lesion had significantly decreased in size. (D, E) Axial (D) and coronal (E) CT scans of case 2 revealed a well-defined, 3.5 × 2 cm mass in the right orbit. (F) Axial CT scan revealed the mass was slightly decreased in size.

paralysis, hoarseness and misswallowing due to right cranial nerve VII, IX, X, XI, XII palsies. CT scans of the head and neck showed an enhanced lesion from the nasopharynx to the parapharyngeal space of the right side with temporal bone destruction (Fig. 1A). On MRI the lesion showed low signal intensity on T1-weighted imaging and high signal intensity on T2-weighted imaging. Contrast-enhanced T1-weighted MRI showed homogeneous enhancement of the lesion (Fig. 1B). The patient underwent an open biopsy through the cervical transparotid approach. However, pathological examination during operation could not reveal whether the specimen was taken from the tumorous lesion or not. An open biopsy of the vertical portion of the facial nerve through the temporal transcortical approach was performed. Hematoxylin-and-eosin-stained sections revealed a proliferation of fibroblastic or myofibroblastic spindle cells admixed with lymphocytes, plasma cells, and histiocytes (Fig. 2A). Bone destruction and neural invasion were also seen (Fig. 2B and C). Neither obstructive phlebitis nor lymphoid follicles with germinal centers were observed with hematoxylin–eosin stain or elastica-van Gieson stain. Immunohistochemically, the spindle cells were focally positive for alpha-smooth muscle actin (SMA) but negative for ALK. The number of IgG4-positive plasma cells and the ratio of IgG4+/IgG+ plasma cells were 279 per high-power field (/HPF) and 25%, respectively (Fig. 2D and E). For analysis of the degree of IgG4+ or IgG+ plasma cells, the areas with the highest density of positive cells were evaluated. These findings indicated IPT/NOS. Systemic corticosteroids (prednisolone, 0.6 mg/kg/day, 30 mg/day, PO) were used and gradually tapered. The size of the main lesion had significantly decreased in size at 5 months after treatment (Fig. 1C). However, symptoms such as hoarseness and dysphagia caused by cranial nerve palsy did not improve.

2.2. Case 2

An 83-year-old female presented with a 2-month history of low vision and pain in her right eye. Her medical history included angina. Physical examination revealed lateral deviation of the right eye. CT scans revealed a well-defined, 3.5 × 2 cm mass in the right orbit (Fig. 1D,E). Endoscopic sinus surgery was performed to obtain a biopsy specimen. Histologic examination revealed the specimen to be an IPT, similar to case 1 (Fig. 2F). Neither obstructive phlebitis nor lymphoid follicles with germinal centers were observed with hematoxylin–eosin stain and elastica-van Gieson stain. Immunohistochemically, the spindle cells were focally positive for alpha-SMA but negative for ALK. The ratio of IgG4+/IgG+ plasma cells was less than 1% (Fig. 2G and H). The diagnosis was IPT/NOS. Systemic corticosteroid therapy could not be performed due to her low activities of daily living. The patient was injected with 0.7 cc of 40 mg/mL triamcinolone acetonide. Two months and seven months later, she was injected again, and the size of the mass slightly decreased (Fig. 1F). The current CT scans showed stable disease for a year with no eye pain. However, the eyesight of the patient was not improved after treatment.

3. Discussion

IPT has been most commonly reported in the lungs, but it is uncommon in the head and neck region. Orbital pseudotumor, most commonly occurring with head and neck lesions, can be localized or diffuse and can affect any portion of the orbit but is typically unilateral rather than bilateral [1]. In contrast, IPT of the parapharyngeal space is extremely rare and only six cases have been previously reported [2–7].

A proper recognition of this entity is clinically important for the determination of a prognosis and a treatment plan. It is difficult to determine whether the diagnosis is IPT without histologic examination. Histologically, IPT consists of a granulomatous lesion characterized by fibroblastic or myofibroblastic proliferation, prominent plasma cells and acute and chronic inflammatory cells. They occasionally mimic malignant disease in their clinical and radiological features due to infiltrative and destructive proliferation. Reactive lesions such as an infectious disease and