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Castleman's disease restricted to the infratemporal fossa

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Abstract
Giant lymph node hyperplasia (Castleman's disease) is usually reported as a solitary mediastinal tumour, although involvement of other anatomical sites and a multicentric form have been reported. We describe a rare case of Castleman's disease due to its localisation (the left infratemporal fossa) and histology (plasma-cell variant). A brief review of the main clinico-histological characteristics of Castleman's disease is also presented.

Key words: Giant lymph node hyperplasia; Head and neck neoplasms

Introduction

Traditional Castleman's disease is a rare clinico-pathological entity characterized by the presence of a single tumour constituted by hyperplasic lymphoid tissue (Castleman and Towne, 1954). More recently, a multicentric form of the disease has been described (Gaba *et al.*, 1978). The unifocal form is usually asymptomatic and has a good prognosis (Keller *et al.*, 1972). Most cases occur as mediastinal tumours, although involvement of other anatomical sites has been reported, the head and neck being the second most common area (Yi *et al.*, 1995). We describe a rare case of unifocal, plasma cellular type of Castleman's disease located in the infratemporal fossa. Head and neck localisations are described and discussed.

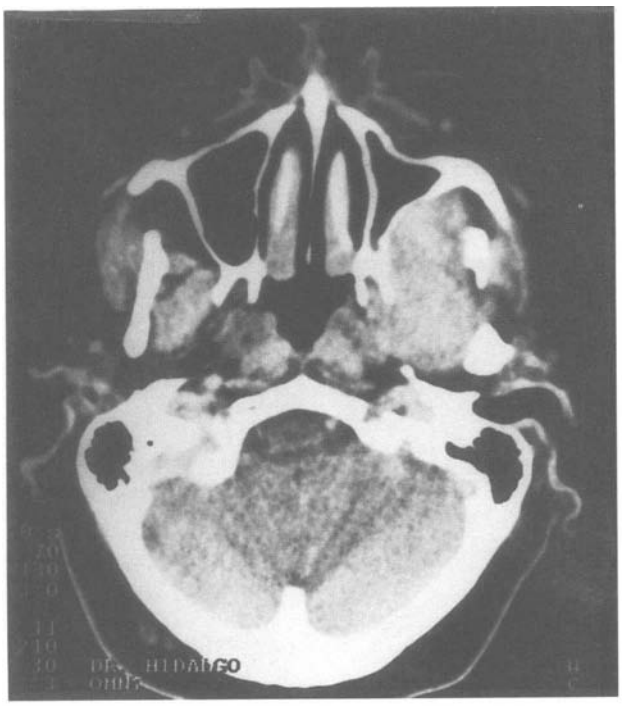

FIG. 1
Axial CT showing a mass in the left infratemporal fossa displacing the posterior wall of the maxillary sinus.

Case report

A 58-year-old woman presented with a two-year history

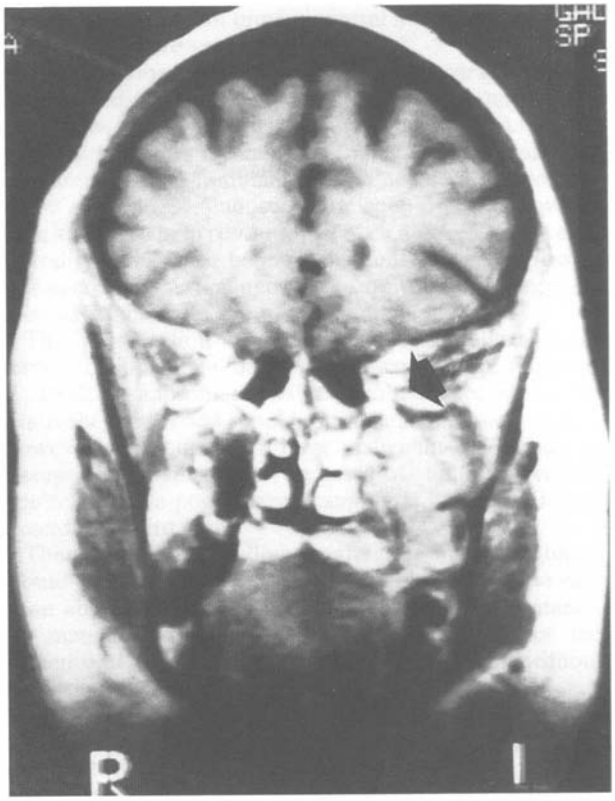

FIG. 2
Coronal contrast-enhanced MRI showing infiltration of nervous and muscular structures by the tumour and extension toward the cranial base through the foramen ovale (arrow).

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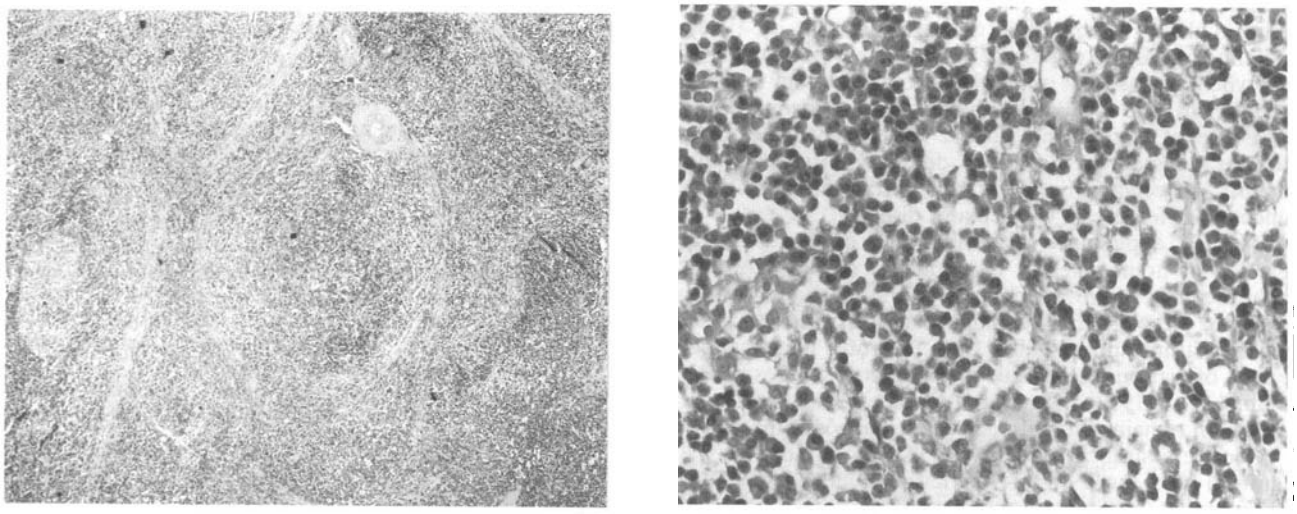

(A) (B)

FIG. 3

Castleman's disease plasma cell type. A, the germinal centre, placed on the centre, is hyperplasic (H & E × 25). B, the cell population between the germinal centres is made of mature plasma cells (H & E × 250).

of painless swelling on the left side of her face associated with hypoesthaesia of this area for the last six months. She denied constitutional symptoms, hoarseness, dysphagia and dyspnoea. The only remarkable finding on physical examination was a firm and non-tender mass under the angle of the mandible. Routine laboratory tests were normal. A computerised tomographic (CT) scan (Figure 1) and a contrast-enhanced magnetic resonance image (MRI) scan (Figure 2) were performed, showing a mass in the infratemporal fossa extending toward the cranial base through the foramen ovale, the pterygo-palatine fossa and displacing the posterior wall of the maxillary sinus. The muscular and nervous structures were invaded by the tumour. The radiological diagnosis was of malignant tumour, but the clinical behaviour belonged to a benign process.

An infratemporal-subtemporal-preauricular approach was performed. The zygomatic process and bone were translocated to access the infratemporal fossa (and repositioned at end of the intervention). A firm mass invading the pterygoidei muscles and the second and third branches of the fifth cranial nerve was identified and excised. The patient had an uneventful post-operative recovery.

Histological examination revealed characteristic features of the plasma cell type of Castleman's disease (Figure 3); hyperplasic germinal centres, not showing hyaline vascular changes, separated by sheets of mature plasma cells and less vascular stroma. Infiltration of muscle and nervous fibres by lymphocytes and plasma cells was also observed. Immunohistochemistry proved the production of Kappa light chains and immunoglobulin G by plasma cells.

Other localisations were excluded after an exhaustive post-operative study. The disease recurred 13 months later in the orbital apex, probably due to progression through the inferior orbital fissure, and it is being controlled with immunosuppressive therapy (prednisone).

Discussion

Since its original description (Castleman and Towne, 1954) Castleman's disease has been reported under a variety of names including giant lymph node hyperplasia, angiofollicular lymph node hyperplasia and angiomatous lymphoid hamartoma. This disease is a benign tumour of lymphoid tissue of unknown aetiology, although the most tenable theories of pathogenesis are that Castleman's disease is either a hamartomatous (Lattes and Pachter, 1962) or inflammatory (Fisher *et al.*, 1970) process.

Two histological variants have been differentiated (Keller *et al.*, 1972): the hyaline vascular type (90 per cent), and the plasma cell type (10 per cent). The hyaline vascular variant consists of small lymphoreticular follicles distributed within a hypervascular hyalinized stroma. The plasma cell variant is composed of larger lymphoreticular nodules separated by sheets of plasma cells and a somewhat less vascular stroma.

The peak incidence of this disease is within the 15-30-year age group, with equal sex prevalence (Keller *et al.*, 1972). Most frequently it shows as an asymptomatic solitary mediastinal mass (70 per cent of cases) (Castleman *et al.*, 1956; Keller *et al.*, 1972) although it has been reported to occur in other anatomical sites including the axilla, retroperitoneum, mesentery, neck, pancreas, muscle, etc. (Cohen, 1957; Daley and Gorog, 1967; Mackay, 1969; Keller *et al.*, 1972; Pujari and Deodhares, 1977). Moreover Castleman's disease may be multifocal (Gaba *et al.*, 1978; Frizzera *et al.*, 1985). Constitutional symptoms (fever, asthenia, weight loss, arthralgia, etc.) and analytical alterations (anaemia, hypergammaglobulinaemia, elevated erythrocyte sedimentation rate, hypoalbuminaemia and elevated alkaline phosphatase) have been associated with 50 per cent of the cases of plasma cell variant and with all cases of the multicentric form (Keller *et al.*, 1972; Frizzera *et al.*, 1985). Non-Hodgkin's lymphomas and Kaposi's sarcoma have been described in association with multicentric type Castleman's disease, but not in association with the localised type.

Castleman's disease is seldom confined to the head and neck, with only 56 cases reported (Yi *et al.*, 1995). Most commonly it appears as an asymptomatic solitary mass in the neck under the border of the sternomastoid muscle (Keller *et al.*, 1972; Pujari and Deodhares, 1977; Gleeson *et al.*, 1988; Rotenberg *et al.*, 1990; Penfold *et al.*, 1991; Sanz *et al.*, 1992; Yi *et al.*, 1995), although it has also been reported to appear in the larynx (Climie *et al.*, 1964), parapharyngeal space (Lanier and Cummings, 1982) and

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parotid gland (Chan and McGuire, 1992; Yi *et al.*, 1995). All the reported cases but one were of the hyaline vascular subtype. Our case was atypical due to its histology (plasma cell variant) and localisation, being, to our knowledge, the first case of Castleman's disease restricted to the infratemporal fossa that has been described. Its infiltrative nature, especially the nervous infiltration, is also an uncommon feature of Castleman's disease.

The CT appearance of Castleman's disease shows a pattern of homogeneous, moderate to marked contrast enhancement, more prominent in the hyaline vascular subtype, although ring enhancement can also be seen, making it indistinguishable from necrotic lymphadenopathy of various neoplastic or infectious causes (Chaloupka *et al.*, 1990). Histopathological examination of the specimen remains the only definitive diagnostic method.

Complete surgical resection is usually curative in the unifocal type, but if part of the lesion is left behind, slow enlargement of the remaining mass is to be expected (Chaloupka *et al.*, 1990; Rotenberg *et al.*, 1990; Penfold *et al.*, 1991). The only head and neck recurrence was reported with the plasma cell type (Sanz *et al.*, 1992). In addition, the infiltrative nature and recurrence of our case, warrants close follow-up after excision of the plasma cell type. Systemic immunosuppressive therapy may be as necessary for the recurrent plasma cell type as for the multicentric type (Sanz *et al.*, 1992).

In conclusion, localized Castleman's disease is a rare benign tumour that occasionally appears as a head and neck mass, and should be considered in the differential diagnosis of any solitary mass in this area. Removal is usually not difficult depending on the site, with a high success rate, although the plasma cell type requires close observation.

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