Castlemans disease in the orbit. A 20-year follow-up

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ABSTRACT.

Purpose: To report a case of localized orbital Castlemans disease of mixed cell type with a follow-up of 20 years.

Methods: A female patient presented at the age of 12 years with constitutional symptoms and left-sided proptosis. Laboratory tests revealed marked hypergammaglobulinaemia and high erythrocyte sedimentation rate (ESR), suggesting an immunological disturbance. A CT scan and MRI showed an infiltrating orbital mass lateral to and behind the eye.

Results: Histological examination of orbital biopsies showed a lymphoid lesion consistent with Castlemans disease of the mixed cell type. The patient was treated with systemic steroids, immunosuppressives and irradiation. She is now 33 years old and has been without relapse for the last 7 years.

Conclusion: Orbital involvement in Castlemans disease is very rare. The clinical course, good prognosis and histological picture of the present case favour the diagnosis of localized Castlemans disease of mixed cell type. The successful medical treatment suggests that such a regime may substitute for surgery when the latter proves difficult.

Key words: Castlemans disease – lymphoid lesion -mixed cell type – orbit

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Castlemans disease (CD) is a rare, atypical lymphoproliferative disorder of uncertain aetiology. Histologically, the disease has been classified into three types: hyaline-vascular, plasma cell, and mixed (transitional) cell type (Keller et al. 1972; Frizzera 1988).

Cases of CD can be clinically divided into two categories. Localized (unicentric) cases usually involve a solitary mass most commonly in the mediastinum. Systemic ( multicentric) cases present with generalized lymphadenopathy and may involve the liver, spleen and other locations (Chen 1984; Frizzera et al. 1985; Weisenburger et al. 1985; Frizzera 1988). Ophthalmic involvement in CD is very rare. Only three cases of ophthalmic involvement are described in the literature (Gittinger 1989; Snead et al. 1993; Kurokawa et al. 1999).

We report a case of orbital CD of mixed cell type with a follow-up period of more than 20 years.

Case Report

Clinical history

The patient described is now aged 33. In 1980, at the age of 12 years, the patient reported repeated episodes of high fever, arthralgia, skin rash and splenomegaly without lymphadenopathy or hepatomegaly. Laboratory tests showed marked hypergammaglobulinaemia and high erythrocyte sedimentation rate (ESR), suggesting an immunological disturbance. The patient responded to acetylsalicylate, with complete remission of the splenomegaly. In 1987, the subject developed an acute left-sided keratoconjunctivitis, with swelling of the lid, restricted ocular motility, side-gaze diplopia and a 6 mm proptosis. Visual acuity (VA) and ophthalmoscopy were normal. A CT scan revealed an orbital mass temporally and behind the eye involving the lateral and superior rectus muscles (Fig. 1A). The patient was treated with systemic prednisone (40 mg/day). In 1988, she reported another attack, this time with an 11 mm proptosis. Magnetic resonance imaging showed progression in the orbital mass and infiltration of the retrobulbar tissue (Fig. 1B). Several biopsies were taken from the left orbit via an inferior orbitotomy. After the histopathological diagnosis was made, the patient was placed on immunosuppressive therapy of prednisone (60 mg/day) and azathioprine (150 mg/day). The left orbit was subsequently irradiated. This treatment resulted in the reduction of the proptosis. Thereafter, the patient had repeated exacerbations that settled down with immunosuppressive therapy. The patient’s condition has been static without any treatment for the last 7 years. The only symptoms are a slight swelling of the lid, a 2–4 mm proptosis and left-sided dry eye.

Histopathology

Two imprints and five biopsies were examined. Microscopically, tissue from the centre of the tumour revealed a hyperplastic lymphoid tissue of mature lymphocytes filling a meshwork of blood vessels. The lymphocytes were arranged in rows and concentric layers around the vessels and invaded their walls, splitting them up like ’onion skin’ (Fig. 2A). Samples from the peripheral parts of the tumour were composed of a high number of mature lymphocytes with many foci of plasma cells and activated lymphocytes (Fig. 2B). No germinal centres were seen.
Fig. 1. (A) A CT-scan of the left orbit (1987), showing a mass occupying the upper and lateral aspects of the orbit (asterisks). (B) An MR scan of the left orbit (1990), showing the orbital mass infiltrating the lateral aspect of the orbit (asterisks).

Fig. 2. (A) Histological section of the orbital biopsy showing the central part of the tumour with features of hyaline-vascular morphology. Note the central hyalinized vessel with concentric rings of lymphocytes, in an 'onion skin'-like arrangement (HE) (lab. no. 493/88c, ×100). (B) Peripheral part of the tumour showing numerous plasma cells (HE) (lab. no. 493/88c, ×100).

Unna-Pappenheim staining confirmed the high number of plasma cells. The majority of lymphocytes reacted with pan-B (CD45R), and demonstrated, like the plasma cells, a polyclonal pattern. The histological picture, along with the clinical presentation, suggested a diagnosis of CD of mixed cell type.

Comments
In CD, the clinical picture, treatment and prognosis are strongly correlated to the histological morphology.

The localized type of CD commonly (90%) presents as the hyaline-vascular type (Keller et al. 1972; Frizzera 1988). Clinically, most cases present as a solitary mass without other symptoms (Frizzera 1988). Rarely, localized CD (10%) is either of pure plasma cell type or of mixed cell type (Flendrig 1970; Keller et al. 1972; Frizzera 1988). The mixed cell type is described as a hyaline-vascular lesion with foci of numerous plasma cells. Both forms of localized CD are amenable to surgical therapy with excellent results and good prognosis (Menke et al. 1992; Chronowski et al. 2001). Radiotherapy may achieve a clinical response in selected cases (Chronowski et al. 2001).

The systemic form of CD is mainly of the plasma cell type and occasionally of the mixed cell type (Menke et al. 1992). Clinically, systemic CD presents with severe generalized symptoms and is often seen in older age groups. Lymphadenopathy is the main presenting sign. Liver and spleen involvement are common (Frizzera et al. 1985; Weisenburger et al. 1985) and prognosis is usually poor.

Castleman’s disease is basically a lymph node disease. Rarely, CD may develop in tissue depleted of lymphatics, like the brain (Severson et al. 1988), and the orbit, as in the present case. Gittinger (1989) reported a case of CD with ocular signs, but no orbital mass was mentioned and no orbital biopsy was taken. Kurokawa et al. (1999) reported a case of systemic CD with an orbital mass. Again, no orbital biopsy was taken. The only case of CD with a histologically proven orbital lesion was a localized, hyaline-vascular type reported by Snead et al. (1993).

In the present case, the patient’s youth at presentation, the absence of lymphadenopathy and the good prognosis (i.e. survival of more than 20 years) favour a diagnosis of localized CD. The splenomegaly was of a transient nature and indicates an immune response rather than a presenting sign of a systemic disease.

The clinical picture and laboratory findings fit the histological diagnosis of the mixed cell type of CD. The good prognosis achieved in this case is probably due to the localized nature of the disease, the absence of generalized lymphadenopathy and hepatomegaly and the polyclonal nature of the disease. The successful treatment with combined therapy of steroid, immunosuppressives and radiotherapy suggests that such a regime may substitute for surgery when the latter proves difficult.

Our case is the first case of localized orbital Castleman’s disease of mixed cell type with a favourable prognosis for more than 20 years to be reported.

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References


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