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Castleman's disease

A 65 year old woman presented in February 1998 with joint pains, mild weight loss, and a low grade irregular fever. Initially, pain was localised around both shoulder joints. Subsequently, elbows, wrists, hips, and knees were affected, with morning stiffness, but without particular swelling. The symptoms were accentuated by movement, but also persisted during the night, often keeping the patient awake. On clinical examination, there was limited painful movement of the shoulders and hips, with a marked reduction in strength. The small joints of the hands and feet were not affected. No other pathological conditions were found. Laboratory findings showed a marked increase in erythrocyte sedimentation rate (ESR; >100 mm/1st h), hyper-α_2 globulinaemia and a mild anaemia, whereas enzymatic activity (serum aspartate aminotransferase, serum alanine aminotransferase, alkaline phosphatase, lactate dehydrogenase, and creatine kinase) was within the normal range. A diagnosis of polymyalgia rheumatica was made and a rapid and marked clinical improvement was obtained with low dose steroid treatment (prednisone 12.5 mg/day). Pain disappeared, muscle strength and joint function became normal within a week. A decrease in ESR (to 40 mm/1st h) and α_2 globulinaemia was noted after one month. The clinical condition remained satisfactory during 1998, with a complete normalisation of ESR and α_2 globulinaemia after three months.

In January 1999 she developed jaundice with pruritus and a subcontinuous fever (38.5–39°C). Axial tomography and nuclear magnetic resonance disclosed compression of the biliary duct by a compact, well defined retropancreatic mass (about 4 cm in diameter). The laboratory pattern was typical of cholestasis (hyperbilirubinaemia, increased serum alkaline phosphatase activity). Endoscopic retrograde colangiopancreatography showed slight enlargement of the biliary tract, dilatation of the gall bladder walls, and absence of gall stones. Transit of contrast medium along the biliary tract slowed down.

Explorative laparotomy confirmed the presence of a mass compressing the common bile duct. This was removed and histological examination disclosed a lymph nodal architecture characterised by evident follicular hyperplasia. Some of the germinal centres were enlarged and comprised polymorphous follicular centre cells, whereas other germinal

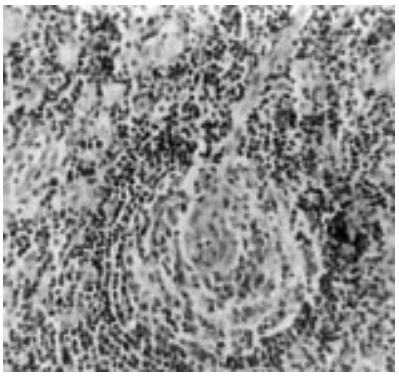

Figure 1 The post-capillary vessels penetrate the expanded follicles perpendicularly ("lollipop image"). (×400.)

centres were depleted of lymphocytes, and consisted predominantly of dendritic reticular cells showing vascular proliferation. The mantle zone was expanded and concentrically arranged around the atrophic germinal centres (onion skin layers). The interfollicular areas were also prominent, containing small lymphocytes, occasional eosinophils, plasma cells, and some immunoblasts, and showing numerous hyperplastic vessels of the post-capillary venous type. Occasionally these vessels, which were often hyalinised, penetrated the expanded follicles perpendicularly, giving rise to the so-called "lollipop" appearance (fig 1). An immunocytochemical study confirmed the normal organisation of the nodal structures, with a clear positivity of follicular elements for typical B cell markers CD20 and CD79a, and positivity of interfollicular lymphoid elements for T cell markers CD3 and CD45R0, whereas the dendritic reticular cells showed a typical positivity for CD21.

These features are distinctive of Castleman's disease, also known by the descriptive term angiofollicular lymph node hyperplasia. This is a clinical entity characterised by angiofollicular hyperplasia of the lymph nodes without the presence of any atypical cells or other signs of malignancy. Many (multicentric or systemic form) or single (monocentric or solitary form) lymph nodal groups can be affected in the process[1] and two histological subsets have been recognised: a hyaline-vascular type, characterised by marked expansion of the mantle follicular zone and a plasma cell type, with diffuse plasma cell proliferation in the interfollicular tissue.[2] The cause of the disease is unknown, but overproduction of interleukin 6[3] has been shown in the course of Castleman's disease and a possible pathogenic role for this cytokine has been suggested. It is commonly associated with several autoimmune conditions such as systemic lupus erythematosus, Behçet's disease, amyloidosis, and with various neoplastic diseases,[4–7] but its onset during the course of polymyalgia rheumatica has not been recently reported. No data exist about the possible relation between Castleman's disease and polymyalgia, but a role for interleukin 6 can be suggested,[8–10] perhaps through a stimulating action of this cytokine on monocytes and lymphocytes. The present case can be considered typical of the solitary form, hyaline-vascular type of Castleman's disease. Our patient did not present any other lymphatic disease during the subsequent months, which would seem to confirm the good prognosis of this disease.

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