Case Report

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Castleman's Disease: An Unusual Cause of a Neck Mass

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Key Words

Lymph node hyperplasia
Castleman's disease
Cervical mass

Abstract

Castleman's disease is an unusual cause of a neck mass; in only 6 % of the cases reported in the literature is the disease primarily located in the neck. We present 2 cases of Castleman's disease restricted to the neck, who underwent successful surgical excision. We discuss the histopathology in the different forms of the disease and review the literature.

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Introduction

Castleman's disease is an uncommon cause of a neck mass [1-7]. Although the head and neck region is the second most common area after the mediastinum, only 57 cases have been reported in the literature [2, 8]. Here we present 2 cases of Castleman's disease as a cause of a solitary neck mass and summarise the salient features of this disease.

Case Reports

Case 1. A 17-year-old female presented to the ENT department with a neck mass which had been present for 1 year. Clinical examination revealed a solitary, firm, 3 cm, non-tender, mobile, left jugulo-digastric node. Panendoscopy was normal and excision biopsy of the neck mass showed

angiofollicular hyperplasia, consistent with Castleman's disease of the hyaline-vascular type (fig. 1). Blood tests including FBC, ESR, serum electrolytes and liver function tests were normal and chest X-ray was clear. The patient was followed up in the outpatient clinic with no further treatment being required and no further lymphadenopathy found after 2-year follow-up.

Case 2. The second patient was a 29-year-old female who presented with a 1 cm mobile mass in the mid-cervical region which had been present for 6 months. Head and neck/ENT examination was otherwise unremarkable. Panendoscopy was normal and the mass

Fig. 1. Angiofollicular lymph node hyperplasia showing onion skin arrangement of lymphocytes around a central hyalinised blood vessel. HE. $\times 200$.

was excised. Histology of the excised lymph node showed typical features of the hyaline vascular variant of angiofollicular hyperplasia (Castleman's disease). Ultrasound of the abdomen and pelvis, chest X-ray, blood tests including FBC, ESR, serum electrolytes and liver function tests were all found to be normal. Follow-up in the outpatient clinic for 3 years confirmed no evidence of recurrence.

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Discussion

Castleman first described a tumour-like mass in the mediastinum which he believed to be lymph node hyperplasia, but not thymic or neoplastic in origin [9]. The peak incidence of the disease is in the 15- to 30-year age group [1]. While in its classical form the disease occurs as a solitary lesion, there have been several reports of patients with multicentric giant lymph node hyperplasia and this latter variant is often seen in patients with acquired immune deficiency syndrome (AIDS) or AIDS-related complex [10].

There are two main histological types: the hyaline-vascular type (80-90%) and the much rarer plasma-cell type (10-20%) [1]. The lesions of the hyaline vascular type consist of small lymphoreticular follicles distributed within a hypervascular hyalinised stroma. Among the vessels is a variable mixture of cells, predominantly lymphocytes which are arranged in 'onion-skin' layers around the follicle centres [1, 8, 11, 12]. The plasma cell variant is characterized by normal to large follicle centres, the presence of sheets of mature plasma cells and somewhat less vascular stroma [1, 8, 11, 12]. A third mixed variant suggesting a transformation of plasma cell into hyaline vascular type has also been described [12].

The hyaline vascular type is usually unaccompanied by systemic manifestations. In contrast the plasma cell variety is usually accompanied by various systemic symptoms; they include fever, anaemia, elevated ESR and altered plasma protein levels [1, 8]. No clear aetiology for Castleman's disease has ever been discussed [1]. The natural history of the disease is continued enlargement of the lymph node with surgical excision being the treatment of choice. It is important to be aware that this rare lesion is benign and, if excision is complete, recurrence is unlikely.

References

Keller AR, Hochholzer L, Castleman B: Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. *Cancer* 1972;29:670-681. Yi AY, de Tar M, Becker TS, Rice DH: Giant lymph node hyperplasia of the head and neck (Castleman's

disease): A report of five cases. *ORL Head Neck Surg* 1995; 113:462-466. Gleeson MJ, Cassidy M, McMullin JP: Castleman's disease – an unusual neck mass. *J Laryngol Otol* 1988;102:661-662. Pujari BD, Deodhare SG: Cervical angiofollicular lymph node hyperplasia (lymphoid hamartoma). *J Laryngol Otol* 1977;91:541-547. Penfold CN, Cottrell BJ, Talbot R: Neonatal giant lymph node hyperplasia (Castleman's disease) presenting in the head and neck. *Br J Oral Maxillofac Surg* 1991;29:110-111.

Kooper DP, Tiwari RM, van der Valk P: Castleman's disease as an uncommon cause of a neck mass. *Eur Arch Otorhinolaryngol* 1994; 251:370-372.

Sanz C, Sierra J, Cobarro J, Avellaneda R, Montserrat E, Rozman C: An unusual case of Castleman's disease restricted to the neck. *ORL* 1992;54:331-333. Rodrigo JP, Fernandez JA, Alvarez JC, Gomez J, Suarez C: Castleman's disease restricted to the infratemporal fossa. *J Laryngol Otol* 1996; 110:896-898.

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Castleman B, Iverson L, Menendez VP: Localized mediastinal lymph-node hyperplasia resembling thymoma. *Cancer* 1956;9:822-830. Lowenthal DA, Filippa DA, Richardson ME, Bertoni M, Straus DJ: Generalized lymphadenopathy with morphological features of Castleman's disease in an HIV-positive man. *Cancer* 1987;60:2454-2458.

Batsakis JG: Tumours of the Head and Neck. Clinical and Pathological Considerations, ed 2. Baltimore, Williams & Wilkins, 1979, p 482. Wolf M, Kessler A, Horovitz A: Benign angiofollicular lymph node hyperplasia (Castleman's disease) presenting as a solitary cervical mass. *J Oral Maxillofac Surg* 1991;49:1129-1131.

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