

Case report

Castleman's disease of the neck: a description of four cases on contrast-enhanced CT

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Abstract. Castleman's disease of the neck is an uncommon benign lymphoproliferative disease that usually presents as homogeneously enhancing enlarged lymph nodes on contrast-enhanced CT scan. We described the appearance of four confirmed cases of Castleman's disease of the neck on contrast-enhanced CT scan. Three of these presented as a solitary enhancing lymph node and the fourth case presented with multiple bilateral enhancing lymph nodes. A central non-enhancing area was present in two of the three cases that presented as a solitary node. Pathological correlation of one of these cases showed that this was due to a central fibrotic scar. One of the enhancing nodes in the fourth case with multiple and bilateral lymphadenopathy also contained a central non-enhancing area. We would like to propose that if a central non-enhancing scar is observed in an enhancing lymph node in the neck on CT scan, Castleman's disease should be considered as a possible diagnosis.

Castleman's disease is an uncommon benign lymphoproliferative disorder that is characterized by hypervascular lymphoid hyperplasia [1, 2]. When this condition affects the neck, it usually presents as a solitary neck mass. There are several previous publications on the imaging features of Castleman's disease of the neck and most of these are single-case reports [1–5]. We would like to describe the appearance of four confirmed cases of Castleman's disease of the neck on post-contrast CT scan, and to suggest that a central non-enhancing scar, if present, is a useful diagnostic clue of the disease.

Materials and methods

Four patients underwent CT scan of the neck as their first line investigation, after presenting with a neck mass. The scan parameters were: 5 mm slice thickness, angle of scan parallel to the hyoid bone, scanning from the level of the external auditory canal to the root of the neck, 20 cm field of view, 135 Kv, 200 mAs and 512 × 512 matrix. All the patients, except in case 2, were given an intravenous bolus dose of 75 ml of non-ionic iodinated contrast agent. The patient in case 2 was given a bolus dose of 50 ml. Scanning for all cases started at 60 s from the onset of contrast injection.

Case 1

A 33-year-old man presented with a painless, non-tender, mobile, level II lymph node of 1 year's duration. Endoscopy of the upper aerodigestive tract was normal. CT scan showed an enhancing (136 Hounsfield units) level

II lymph node measuring 4.5 cm in the longest diameter on the left side of the neck. A low-attenuation crescentic band was observed in the centre of the mass (Figure 1). The mass was excised and histopathological evaluation revealed Castleman's disease of the hyaline vascular type.

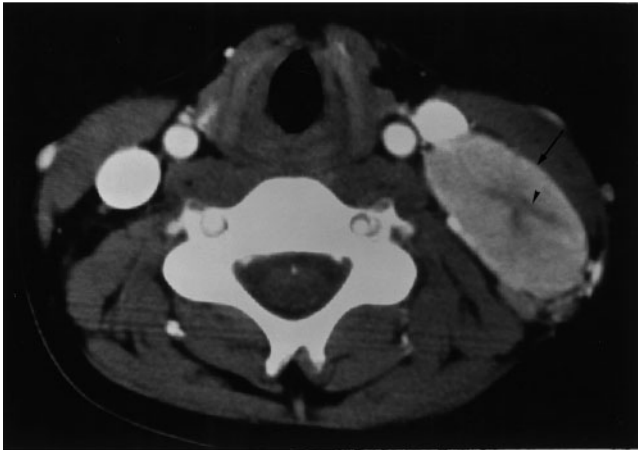
Case 2

A 12-year-old boy presented with a 4 cm non-tender, mobile level III neck lymph node on the left side for a duration of 3 months. Nasopharyngoscopy showed a normal upper aerodigestive tract. CT scan showed a left-sided 4 cm ovoid enhancing (140 Hounsfield units) lymph

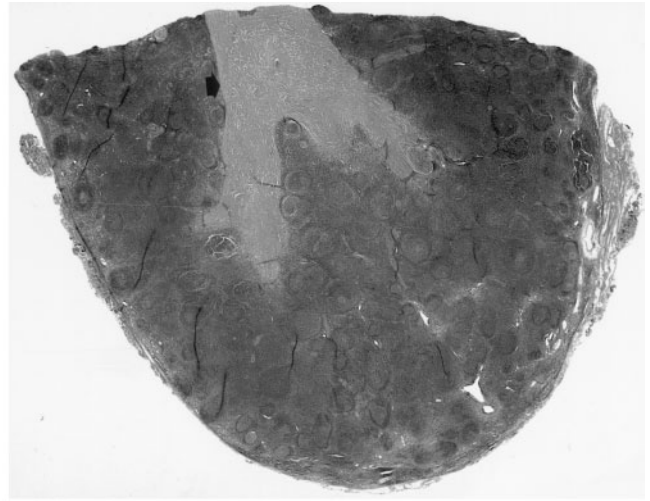


Figure 1. Axial contrast-enhanced CT scan of the neck showing the enlarged and enhancing Level II lymph node on the left side (white arrow) with a crescentic low-attenuation band in the centre (arrowhead).

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(a)



(b)

Figure 2. (a) Axial contrast-enhanced CT scan of the neck shows the enlarged enhancing left Level III lymph node (arrow) with low-attenuation stellate bands forming a scar in the centre (arrowhead). (b) Hemisectioned lymph node showing part of the central stellate fibrous scar (broad arrow). Haematoxylin & eosin stain; original magnification $\times 10$.

node at level III. A low-attenuation stellate band was observed in the centre of the mass (Figure 2a). Histology revealed Castleman's disease of the hyaline vascular type. The gross specimen contained a dense fibrous stroma in a stellate pattern, forming a scar in the centre of the mass (Figure 2b). This correlated very well with the low-attenuation stellate band seen on the CT scan.

Case 3

A 16-year-old girl complained of a painless 2.5 cm lump over the angle of the jaw on the left side for the duration of 2 years. CT scan showed a 3.0 cm homogeneously enhancing mass (154 Hounsfield units) with well-circumscribed margin in the superficial lobe of the left parotid gland (Figure 3). The patient underwent a left superficial parotidectomy and histopathological evaluation revealed hyaline vascular type Castleman's disease.



Figure 3. Axial contrast-enhanced CT scan showing a bright homogeneously enhancing mass in the left parotid gland (white arrow).

Case 4

A 71-year-old man presented with a 2.5 cm non-tender right neck lump for a duration of 1 month. There were no constitutional symptoms. There was no abnormality noted in the upper aerodigestive tract. CT scan revealed multiple enhancing lymph nodes (110–120 Hounsfield units) in both sides of the neck. Their sizes ranged from 1.0 cm to 2.0 cm. Most of these nodes were homogeneously dense although one of the nodes on the right side had a central rounded non-enhancing area (Figure 4). Excision biopsy of one of the right-sided nodes revealed Castleman's disease of the plasma cell type.

Discussion

Castleman's disease is an uncommon lymphoproliferative disorder with a characteristic hypervascular lymphoid hyperplasia [1, 2]. First described by Castleman and associates in 1956 [6], it has since come with many synonyms. These include angiofollicular mediastinal lymph node hyperplasia, angiomatous lymphoid hamartoma, lymph nodal hamartoma, follicular lymphoreticuloma and benign giant lymphoma [7]. The many synonyms reflect the uncertainty over its pathogenesis, although most authors would regard Castleman's disease as a hamartoma or an inflammatory/infective lesion [2, 8].

Histologically, the disease can be divided into two types; the hyaline vascular type and the plasma cell type. The hyaline vascular type makes up about 90% of the cases [1]. Microscopically, the hyaline vascular type is characterized by abnormal small follicles and interfollicular vascularity; consisting of a network of small capillaries with thickened, hyalinized walls radially penetrating the germinal centres from the perifollicular tissue. Follicles characterized by concentric layering of lymphocytes within germinal centres can also be seen. Large fibrotic masses surrounding vessels are often found scattered in the interfollicular areas [7]. The plasma cell type makes up about 10% of the cases [1]. It is characterized microscopically by solid sheets of plasma cells in the interfollicular area. The follicles are

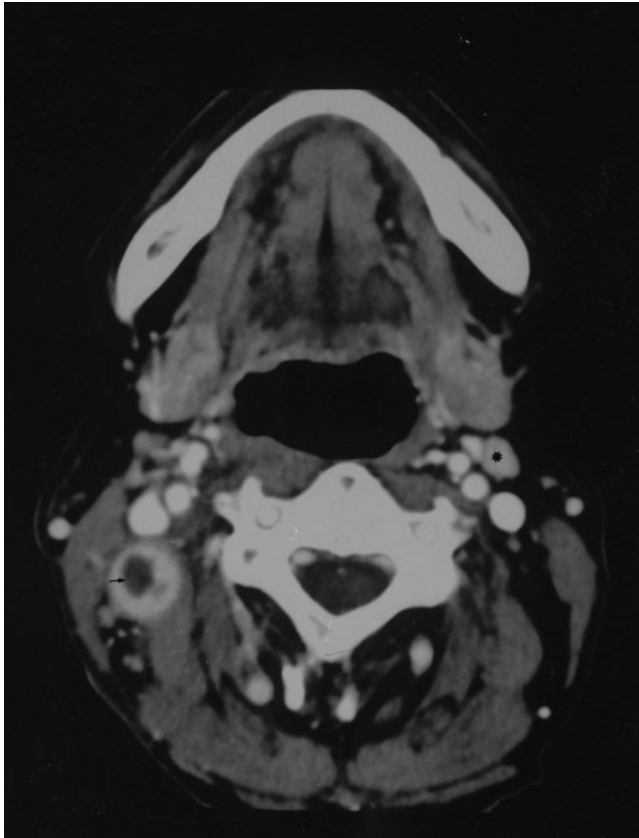


Figure 4. Axial contrast-enhanced CT scan showing a right sided enhancing lymph node with a central rounded non-enhancing area (small arrow). A homogeneously enhancing lymph node is also noted on the left side (*).

usually larger and the prominent interfollicular capillary network characteristic of the hyaline vascular type is usually lacking in the plasma cell type [7].

More than 50% of patients with plasma cell type of Castleman's disease have systemic manifestations including fever, fatigue, anaemia, hyperglobulinaemia and elevated sedimentation rate. Our patient in case 4 though, did not have any constitutional symptoms. Only about 3% of patients with hyaline vascular type of Castleman's disease exhibit these features [8]. Most patients with hyaline vascular type of disease are asymptomatic, although they can present with compressive symptoms caused by the mass lesion [1]. Castleman's disease has no sex predilection and the age of presentation ranged from 8 years to 70 years, although the youngest patient reported was diagnosed at 6 weeks after birth [8].

Surgical excision is the treatment of choice for Castleman's disease of the neck. There is no reported recurrence for the hyaline vascular type. However, plasma cell type requires closer follow-up after the surgical excision and systemic chemotherapy may be required [8].

Castleman's disease is usually limited to one site, although a widespread and aggressive form involving lymphadenopathy in several sites with splenomegaly has also been described [9]. The most common site of the localized form is the mediastinum (about 70% of cases). The neck is the next most common [1, 4]. Most of the previously reported cases of Castleman's disease of the neck were of the hyaline vascular type and these usually present as a

solitary mass lesion both on clinical examination and on imaging [1–5, 8]. This was also the observation in our cases, as the patients in case 1 to 3 all presented with solitary neck masses on imaging and were all of the hyaline vascular type. The exception was in case 4 where the patient presented with multiple bilateral lymphadenopathy and the histology was of the plasma cell type. It may be that plasma cell type of Castleman's disease of the neck has a greater tendency to present as multiple rather than solitary neck masses, although a further study with a larger number of cases would be needed before this impression could be confirmed.

Castleman's disease of the neck on CT scan has been described as well-circumscribed homogeneous mass lesion with moderate to intense enhancement [1, 3–5, 8]; with the hyaline vascular type having a tendency to enhance more than the plasma cell type, due to the greater vascularity of the former [2, 10]. One case of Castleman's disease of the neck presenting with ring-enhancement on CT scan has also been described [2]. Unlike pelvic disease where calcification can occur in up to 50% of the cases [10, 11], calcification in the neck disease is uncommon. On MRI of Castleman's disease of the neck, some authors have described the presence of linear hypointense signals in a stellate or arborizing pattern especially on the T_2 weighted sequences [1, 4, 12]. They attributed these hypointense signals to perivascular lamellar fibrosis or sinus histiocytes and radial fibrosis; and suggested that these hypointense signals could be an important diagnostic clue of Castleman's disease [1, 4, 12]. In contrast, the appearance of Castleman's disease of the neck on CT scan has often been described as non-diagnostic [4, 8]. This is because other disease conditions like lymphoma, tuberculosis, metastatic papillary thyroid carcinoma, Kaposi's sarcoma and Kimura's disease can also present with enhancing lymph nodes in the neck [13]. Ota and co-authors reported on a case of hyaline vascular Castleman's disease of the abdomen which had a central stellate fibrotic area within the mass on CT scan [14]. Similar changes have not been described before in previous reports of CT appearance of Castleman's disease of the neck. Crescentic, stellate and rounded areas of non-enhancement were observed in three of our four patients (cases 1, 2 and 4, respectively). Correlation with the gross specimen in case 2 showed that dense fibrous stroma forming a scar was responsible for the appearance, similar to what was reported in Ota's paper. Although no correlation was made with the gross specimen in cases 1 and 4, we believe that dense fibrous scar could also be responsible for the crescentic and rounded areas of non-enhancement in these two cases, respectively. The absence of a central non-enhancing area in the intraparotid mass of case 3 could possibly be due to an absence of a dense concentration of fibrous tissue. We would therefore like to propose that the presence of a central non-enhancing scar in an enhancing lymph node in the neck on contrast-enhanced CT scan could be an important diagnostic clue of Castleman's disease.

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