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Doppler Findings In Castleman Disease - A Rare Case

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Abstract

Castleman's disease is a rare, benign disease of unknown cause that induces reactive lymph node hyperplasia. It has two histologic subtypes: hyaline vascular and plasma cellular. A definitive diagnosis necessitates tissue biopsy. A specimen may be even misdiagnosed as lymphoma in frozen section. Surgery is the treatment of choice for the solitary form, whereas chemotherapy, radiotherapy, and steroids are proposed for the multicentric type. When this condition affects the neck, it usually presents as a solitary neck mass. Multiple neck masses are an uncommon presentation of Castleman disease affecting the neck. Most Castleman's disease lesions appear as nonspecific, well-defined hypoechoic masses on sonography. Sonography remains useful for the evaluation of cervical and axillary Castleman's diseases, in which the depiction of prominent peripheral vessels and penetrating feeding vessels on Doppler sonograms can suggest the diagnosis of this uncommon disease. Castleman's disease of the neck on CT and MRI scan has been described as well-circumscribed homogeneous mass lesion with moderate to intense enhancement.

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Key words : - Castleman's disease, doppler

INTRODUCTION

Castleman's disease is a rare, benign disease of unknown cause that induces reactive lymph node hyperplasia, most commonly in the mediastinum. It has two histologic subtypes: hyaline vascular and plasma cellular. A definitive diagnosis necessitates tissue biopsy. A specimen may be even misdiagnosed as lymphoma in frozen section. Surgery is the treatment of choice for the solitary form, whereas chemotherapy, radiotherapy, and steroids are proposed for the multicentric type.

We will like to report the sonographic and Doppler findings of Castleman disease of neck in a 13 year old boy. When this condition affects the neck, it usually presents as a solitary neck mass. Multiple neck masses are an uncommon presentation of Castleman disease affecting the neck. There are several previous publications on the imaging features of Castleman's disease of the neck and most of these are single-case reports with emphasis on CT and MRI findings [1]. Konno et al. [2] have described Doppler findings in a case of mesenteric Castleman's disease Doppler findings in a mesenteric node. To best of our knowledge, colour Doppler findings in a Castleman disease of neck has not been reported in the literature as a case report.

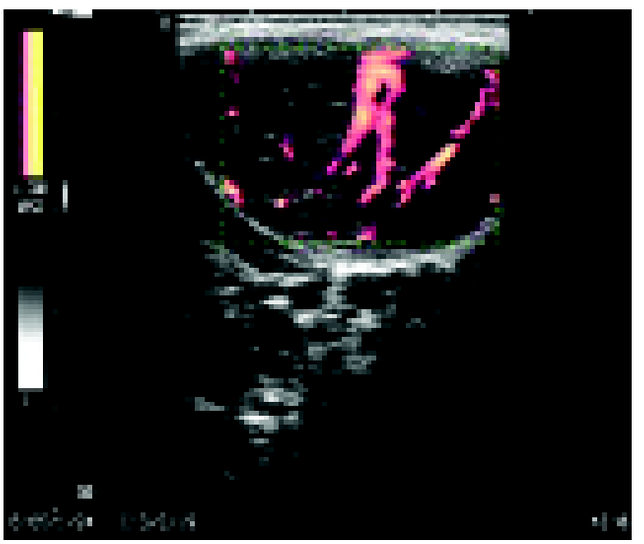

FIG.1 Power doppler image of enlarged left submandibular region lymphnode shows large feeding artery penetrating the nodal hilum and prominent vascular channels in the periphery and the center of the lesions.

CASE REPORT

A 13 years old boy presented to us with complains of a painless swelling in neck for one year. Ultrasound examination of the neck was performed. B-mode

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ultrasound showed multiple well-defined, hypoechoic lymph nodes in left submandibular region, right submandibular region and middle jugular group of nodes. The largest lesion was seen in the left submandibular region measuring 3.5 cm x 2.3 cm in largest dimensions. No evidence of any calcification or necrotic area was seen within the lesion. On color and power mode the lesion was highly vascular. A large feeding artery penetrating the nodal hilum and prominent vascular channels in the periphery and the center of the lesions were identified on Doppler sonogram. Spectral waveform shows both arterial and venous type of pulse wave pattern. In view of multiple well-defined hypoechoic hypervascular lesions, two possibilities were kept- lymphoma and Castleman disease.

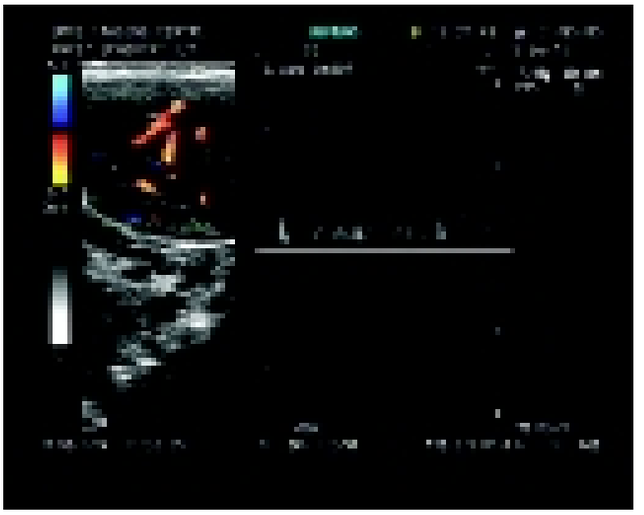

FIG. 2A Color and power mode image of enlarged neck lymphnode shows increased vascularity of lymphnode with arterial waveforms on spectral analysis.

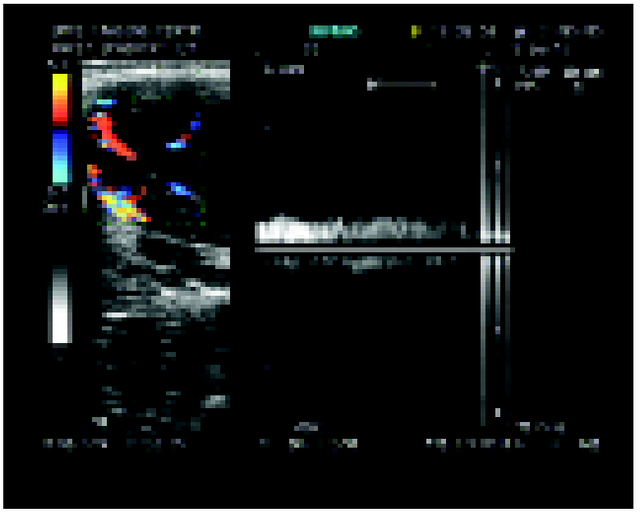

FIG. 2B Color and power mode image of enlarged neck lymphnode shows increased vascularity of lymphnode with venous waveforms on spectral analysis.

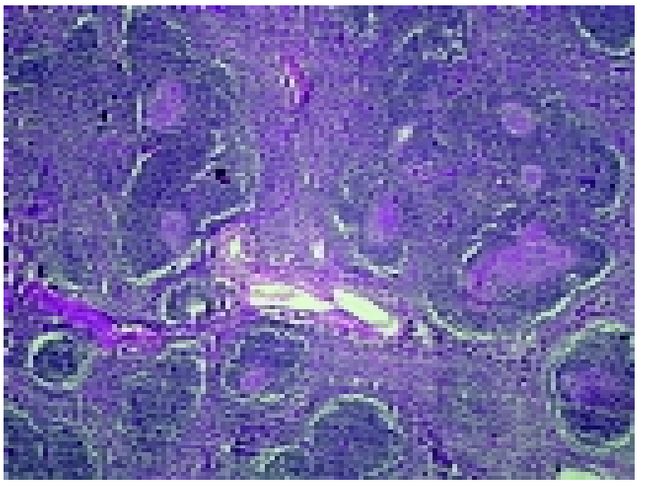

FIG. 3 The section shows follicles with marked vascular proliferation and hyalinization of their abnormal germinal centers. Interfollicular stroma is prominent and shows numerous hyperplastic vessels. The sections from the larger node show the structure of Castleman's disease (also known as giant lymph node hyperplasia, angiofollicular hyperplasia, lymph nodal hamartoma).

Chest x-ray shows no evidence of mediastinal or hilar lymphadenopathy. Ultrasound of abdomen reveals no evidence of mesenteric or retroperitoneal lymphadenopathy.

Excisional biopsy of the lesion in the submandibular region was suggestive of changes of hyaline vascular type of angiofollicular hyperplasia (Castleman's disease).

DISCUSSION

Castleman's disease is an uncommon benign lymphoproliferative disorder, first described by Castleman and associates in 1956 and characterized by hypervascular lymphoid hyperplasia [3]. The cause of Castleman's disease is uncertain; it is thought to be inflammatory or hamartomatous in nature [4]. When this condition affects the neck, it usually presents as a solitary neck mass. Castleman's disease is very rare pathology with unknown precise incidence. More than 400 cases of isolated case are reported in literature so far [3]. The multicentric or systemic form is less common.

Two distinct histologic variants are recognized [5]. The most common is the hyaline-vascular type (more than 90% of the cases), which consists of small lymphoreticular follicles distributed within a hypervascular hyalinized stroma. The plasma cell type is less common (fewer than 10% of the cases) and consists of larger lymphoreticular

nodules that are separated by sheets of plasma cells and a somewhat less vascular stroma. Castleman's disease occurs at any age, with a peak incidence in the third to fourth decades of life, but the multicentric form usually affects older individuals. Both types have equal male female incidence [6]. Patients with the hyaline-vascular-type disease are usually asymptomatic, but they may complain of symptoms caused by the compression of adjacent structures or may present with a palpable mass [7]. In this type usually mediastinal and cervical lymph nodes are involved [5]. Plasma cell type is having typically more generalized (thoracic, mesenteric, and retroperitoneal) lymph node involvement (although localized nodal involvement can be seen) and the disorder tends to be multicentric (although focal disease can be found in 10% of cases). Systemic manifestations are commonly seen in the plasma cell type and include fever, anemia, and hyperglobulinemia [7]. In this type retroperitoneal and mesenteric lymph nodes are mainly involved [5].

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. The most common site of the localized form is the mediastinum (about 70% of cases) followed by the neck [1]. Less than 10% of cases arise in the head and neck [8, 9]. 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]. Additional sites of occurrence include the axilla, retroperitoneum, mesentery, vulva; pancreas and pelvis [7].

Most Castleman's disease lesions appear as nonspecific, well-defined hypoechoic masses on sonography. Konno et al. have described a case of mesenteric Castleman's disease characterized by a large feeding artery penetrating the nodal hilum and prominent arteries in the periphery identified on a Doppler sonogram [2]. Sonography remains useful for the evaluation of cervical and axillary Castleman's diseases, in which the depiction of prominent peripheral vessels and penetrating feeding vessels on Doppler sonograms can suggest the diagnosis of this uncommon disease [6]. Castleman's disease of the neck on CT scan has been described as well-circumscribed homogeneous mass lesion with moderate to intense enhancement; with the hyaline vascular type having a tendency to enhance more than the plasma cell type, due to the greater vascularity of the former. Unlike pelvic disease where calcification can occur in up to 50% of the cases, calcification in the neck disease is uncommon. 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 [1]. 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 T2 weighted sequences. 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].

CONCLUSION

Although most Castleman's disease lesions typically appear as well-defined hypoechoic neck masses on ultrasound, the findings are non-specific. Identification of the large feeding artery penetrating the nodal hilum, hypervascular lymph node and presence of both arterial and venous waveform pattern is an important Doppler characteristic for the diagnosis of this uncommon disease entity.

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