

Unicentric Castleman's Disease: Unusual Disorder of the Neck a Case Review

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ABSTRACT

Castleman's disease (giant or angio follicular lymph node hyperplasia) was first described by Benjamin Castleman in the year 1957 and was named after him. It is an uncommon lympho proliferative disorder which is localised to single lymphnode (unicentric) or multiple lymph node level systemically (multicentric). It is a very rare disorder characterised by non-cancerous growths. The most common sites of this are mainly thorax (mediastinum or lung hilum) and abdomen. It rarely occurs in cervical area.

We report a case of unicentric cervical castleman's disease of neck in an 18-year-old female, who came to ENT OPD with history of right sided neck mass since 6-8 months. After thorough clinical examination and investigations, complete excision of the mass was done. Histopathological examination confirmed the diagnosis of Castleman's disease. On follow-up for one year no recurrence has been seen.

Keywords: Angiofollicular lymph node hyperplasia, Cervical area, Multicentric (MCD), Unicentric (UCD)

CASE REPORT

We report a case of 18-year-old female patient who came to ENT OPD with history of right sided slowly progressing neck swelling since 6-8 months. There was no history of fever or weight loss except mild fatigue. On examination firm, ovoid, non tender and mobile mass measuring 4 × 2 × 5 cm was situated in the right side of the neck. ENT examination and other systemic examinations revealed normal findings. Fine needle aspiration cytology (FNAC) showed moderate cellularity comprising of small lymphocytes in dys-cohesive and in small clusters. These clusters are composed of follicular dendritic cells and occasional spindle cells. These follicular dendritic cells have oval nuclei, evenly distributed chromatin, small nucleoli with abundant delicate cytoplasm containing small lymphocytes along with few plasmacytoid cells. Computed tomography showed intensely homogeneously enhancing well defined lesion located postero-medially to the sternocleidomastoid and postero-laterally to the carotid vessels in the mid portion of the neck (probably carotid space) on right side. USG abdomen and X-Ray chest were done to rule out multicentric type of disease. Hence, diagnosis of Castleman's disease was made by FNAC of the mass and CT scan of the neck [Table/Fig-1].



[Table/Fig-1]: CT scan showing intensely homogeneous enhancing well defined lesion located posteromedially to the sternocleidomastoid and posterolaterally to the carotid vessels in the mid portion of the neck on right side



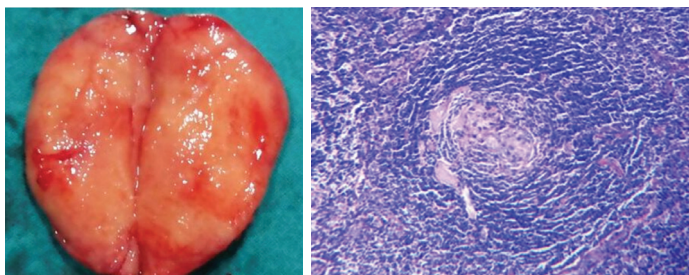
[Table/Fig-2]: Intraoperative image of the tumour

It was decided to excise the mass. Intra-operatively [Table/Fig-2] an ovoid mass was lying in between the carotid artery and internal jugular vein which was meticulously dissected out in toto and sent for histopathological examination.

On gross examination specimen consists of single grey white soft tissue bit measuring 5.5 × 2 × 1.5cm. external surface was glistening [Table/Fig-3]. Section studied shows lymphnode with lymphoid follicles having central prominent blood vessels lined by endothelial cells with hyalinised walls. The lymphocytes surrounding the central vessels are arranged in a concentric fashion. The interfollicular area shows proliferating small blood vessels surrounded by collagen fibres [Table/Fig-4]. It was confirmed as hyaline-vascular type of Castleman's disease. Follow up for one year has been done and no recurrence of mass is seen and patient is relieved of the symptoms.

DISCUSSION

Castleman's disease (CD) was originally identified by Benjamin Castleman in the year 1954 [1], who described a cohort of patients with solitary hyperplastic mediastinal lymph nodes which demonstrated small, hyalinized follicles and interfollicular vascular proliferation on histopathology. Years later, Castleman and



[Table/Fig-3]: Cut section of the specimen **[Table/Fig-4]:** Showing hyaline vascular type of Castleman's disease (10x)

colleagues distinguished this hyaline vascular type of CD from other variants of this disorder, namely the plasma cell variant and the much rarer mixed variant [2]. CD is a disease process of complex array of symptoms, signs and treatment options. It is classified as unicentric and multicentric types. The unicentric variant typically involves single lymphnode while the other type can affect any of the reticulo endothelial organ with involvement of multiple lymph nodes. Unicentric Castleman's disease (UCD) usually presents in young adults with localized masses in the mediastinum (60-75%), neck (20%) or less commonly intra-abdominal masses (10%). Multicentric Castleman's diseases (MCD) are less common than the localized variant.

Histologically, it is classified into 3 variants; a: hyaline-vascular type, b: plasma cellular type and c: mixed type. Amongst these hyaline vascular comprises of 91% [2].

CD affects male and female equally with age incidence ranging from 8-66 y. Several immunological mechanisms have been proposed including over production of interleukin-6 and human herpes virus type -8 infections [2,3]. UCD patients are usually asymptomatic and they have an indolent, slow progressive course, whereas MCD has very aggressive course involving multiple group of lymph nodes and presents with systemic symptoms.

Although CD is a benign disease of the lymphatic's but a literature from France shows that 8 patients has concurrent malignant lymphoma out of which,6 with B-cell NHL and 2 with hodgkins lymphoma. The pathophysiology between the UCD and lymphoma is uncertain. Rajeshwar KV et al., has described that the symptoms of Hodgkin's disease and lymphoma can mimic those of Castleman's disease. In Hodgkin's disease, the symptoms may include fever, night sweats, weight loss, and/or enlarged lymph nodes. The tumours occur most often in the chest, stomach, or the spleen. Hodgkin's disease is usually progressive and it may spread to involve the lymph nodes which are located in other areas of the body [4].

Interestingly, HIV status may influence the constellation of signs and symptoms in MCD patients: based on epidemiologic data from the pre- and post-HIV eras, HIV-negative individuals may take years to manifest any signs or symptoms of the disease, while HIV-positive patients may rapidly develop symptoms over a period of months [5]. The diagnosis is done by histology thereby requiring either biopsy or resection of the mass.

M Sumi et al., has described CT findings of non contrast studies, which showed homogenous mass of soft tissue attenuation in unicentric HV type, Strip type of enhancement was found after a bolus injection of contrast material and rim type on a 5-min delay. Calcification is rarely seen in about 5-10% of the cases, which is coarse and centrally located [6].

The treatment options for UCD are mainly surgical resection of the abnormal node [7]. In patients where the lesion cannot be fully resected, the partially resected mass may simply be observed for progression [3,7]. But for progressing masses 30 to 45 Gy external beam radiation can be considered [3].

On reviewing the literature we found that Vikasdeep Goyal et al., described similar case lymphoid hyperplasia situated in the mesenteric lymphnode associated with AV fistula and anaemia whereas in our case the cervical group of lymphnode was involved with few pressure symptoms and without anaemia [8]. Only few cases of cervical Castleman's disease have been reported in the literature and most of them presented as a solitary neck mass [9].

CONCLUSION

As an ENT and Head and Neck surgeons, we come across many cervical masses in day-to-day practice, although CD is a rare disorder it has to be kept as one of the differential diagnosis of cervical mass. In addition we want to emphasize that approach for MCD and UCD are totally different and surgery remains as the mainstay of treatment for UCD with long term follow-up is necessary as risk of malignancy exists.

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