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A RARE CASE OF UNILATERAL LYMPHADENOPATHY: UNICENTRIC CASTLEMAN DISEASE

Histopathology

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ABSTRACT

Unicentric castleman disease is rare benign lymphoproliferative disorder of unknown etiology. The most common site is mediastinum followed by head and neck, pelvis and retroperitoneum. Only few cases have been reported from outside Indian Subcontinent so far in paediatric age group (≤18 years), presenting with cervical adenopathy. A 15 years female complaining of swelling over left supraclavicular region since birth, increasing progressively in size since past 1 year came to OPD. Fine needle aspiration was suggestive of reactive hyperplasia. Excised enlarged left cervical lymph node revealed hyaline variant of castleman disease. Histopathological diagnosis was confirmed by immunohistochemistry. No recurrence occurred during follow up.

KEYWORDS

Lymph node, lymphoma, lymphoproliferative

INTRODUCTION

Unicentric castleman disease (UCD) rare benign lymphoproliferative disorder of unknown etiology.

Usually asymptomatic Survival rate is 95.3 %. Most common site is mediastinum followed by head and neck, pelvis and retroperitoneum.

Case Study

A 16 years female brought to OPD by relatives with swelling over left supraclavicular region since birth, gradually increasing in size as patient grew in age and associated with pain since past 6 months. Patient had difficulty in walking with discomfort in lower back since many years.

On examination, left cervical region showed approx. 4x4x3 cm sized tender, firm, mobile swelling. Personal history, family history and pat history of patient was found to be non-contributory. Oral examination showed no tonsillar enlargement or dental caries. She had no pallor, icterus, cyanosis, clubbing and or lymphadenopathy elsewhere. Per abdomen examination showed no organomegaly. Systemic examination was found to be unremarkable.

Differential diagnosis like tubercular lymphadenopathy, bacterial lymphadenopathy, malignant lymphadenopathy, and HIV were considered.Patient underwent detailed clinical, radiological and haematological work-up. Patient was prescribed with multivitamins, supplements and antibiotics which do not showed any relief from gradually increasing left cervical swelling associated with pain.

Henceforth, patient underwent local ultrasonography, suggestive of few well defined enlarged heterogeneously hypoechoic necrotic lymph nodes showing internal vascularity in left cervical level II measuring 3.6x2.1 cm. Chest X-ray showed soft tissue swelling in left cervical region [Fig./ Table-1a].

Patient was referred to chest and TB department for tuberculosis workup. Sputum gene xpert was found to be negative. Mantoux test and gastric aspirate for acid fast bacilli were normal. Electro-cardiogram showed sinus bradycardia with sinus arrythmia. Ultrasonography of abdomen and pelvis showed right ovarian haemorrhagic cyst. Complete blood cell count, serum electrolytes, liver function test and kidney function test were found to be within normal limit. Peripheral smear was unremarkable. Triple H status was found to be negative. Contrast enhanced computed tomography of thorax showed multiple enlarged reactive lymph nodes in left cervical level III, IV and V, of which largest measuring 4.2x2.7 cm at level V, suspicious of lymphoma. No evidence of thoracic or abdominal lymphadenopathy [Fig./ Table-1b].

Patient underwent fine needle aspiration of her lump in private OPD which showed scantly cellular smears comprised of moderate inflammatory cell infiltrates predominately comprised of lymphocytes against haemorrhagic background, suggestive of cystic swelling with chronic inflammation. Repeat fine needle aspiration cytology of left cervical lymph node was done in our OPD, suggestive of reactive hyperplasia comprised of polymorphous population of lymphoid cells. No granuloma/ atypical cells seen [Fig./ Table-2a-b].

Patient was admitted under general surgery department for surgical excision of lymph node in view of increasing size. In lymph node gene xpert study, mycobacterium tubercule bacilli was not detected. Enlarged lymph node was excised and sent for HPR. Formalin fixed specimen of left cervical lymph node was received to histopathology department as one large nodal mass measuring 4x3x2 cm. Externally, brown in color with focally intact capsule. Cut surface was found to be pale, whitish, homogeneous, soft in consistency. Microscopic examination of well fixed, prepared and hematoxylin and eosin stained slides serial and deeper sections showed well preserved architecture of lymph node with increased number of lymphoid follicles, scattered throughout the cortex and medulla. Twining and coalescing of germinal centre in many follicles [Fig./ Table-3a]. Prominent hyaline deposits along with sclerotic blood vessels piercing the germinal centres radially [Fig./ Table-3b-c]. Dendritic cell in germinal centre showed proliferation. Focal prominence of mantle zone due to concentric ring arrangement of lymphocytes [Fig./ Table-3d]. Interfollicular regions were composed of numerous vascular channels with plump endothelial cells [Fig./ Table-3e]. No evidence of prominent eosinophils, plasma cells or Reed-Sternberg cells. Congo red stain and acid fast bacilli stain were found to be negative. Correlating clinico-radiological features with cytomorphological and histopathological features were in favour of hyaline vascular variant of castleman disease. Immunohistochemistry is essential to rule out other causes of lymph node enlargement including lymphoproliferative disorder. Immunohistochemistry performed over above sections for

CD23 revealed prominent dendritic cells [Fig./ Table-4a-b]. Germinal centres were BCL-2 negative, confirming their reactive nature [Fig./ Table-4c].

At present, she is under follow up. Patient is hemodynamically stable with no active complaints. She is not on any medication.

DISCUSSION

It is a benign lymphoproliferative disorder of lymphatic system, first described by Benjamin Castleman in 1954 [1]. Its prevalence is 1 in 100,000 cases commonly occurring in adults than paediatric age group (< 18 years) [2]. In children, most common site is thorax [3]. Other sites involved in children in decreasing order of frequency are: abdomen, neck, axilla [4]. It is subtyped as unicentric and multicentric. Histologically, further classified as hyaline vascular, plasma cell type and mixed type [5]. It is a rare diagnosis in paediatric age group. The common causes of lymphadenopathy like infection, tuberculosis, malignancy, were ruled in present case with detailed investigations. Diagnosis was established with histological findings as depicted in [Fig/ Table-3a-e], confirmed on immunohistochemistry as showed in [Fig./ Table-4a-c].

Preexisting literature was reviewed using PubMed, Google scholar. Present study findings were compared with other similar studies done in paediatric age group. Age of presentation was 16 years in present study which was found to be in concordance with study undertaken by Melkundi RS et al. [6] where age of presentation was 18 years. Present study includes female patient which is found to be congruent to previous studies done by Melkundi RS et al., Varma S et al., Ankur Singh et al. [6,7,8]. Present study shows gender incongruence with studies performed by R RS et al., Kansara AH et al., Kumar KM et al., Kausar Z et al. [9,10,11,12] where male patients were affected.

This discordance may be due to few cases reported in literature and or gender discrimination leading to reporting of male patients more as compared to female patients. Present study shows similar mode of presentation as enlarging neck mass, cervical region as site and size ranging in size from 2x2 cm to 7x4 cm when compared with studies carried out by Melkundi RS et al., Varma S et al., Ankur Singh et al., R RS et al., Kansara AH et al., Kumar KM et al., Kausar Z et al. [6,7,8,9,10,11,12]. Left side of neck was affected in present study and studies done by Kansara AH et al. [10]. There is difference in laterality with respect to studies done by Melkundi RS et al., Varma S et al., R RS et al., Kumar KM et al., Kausar Z et al. [6,7,9,11,12].

Both sides of neck were affected in study done by Ankur Singh et al. [8]. Ultrasonography was done multiple times and contrast enhanced computed tomography once in present study which was found to be in concordance with studies carried out by Kumar KM et al. and Melkundi RS et al. respectively [11,6]. Choice of imaging modality purely depends on various factors like list of differential diagnosis, availability. FNAC was done in five out of seven case studies which were found to be non-specific, including present study [6,8,9,11,12]. FNAC plays limited role in approaching these kinds of cases.

Histopathological examination was performed in all cases including present study with exception of study done by Ankur Singh et al. [8], where patient was managed conservatively. The most common variant was found to be hyaline vascular in present study and studies done by Melkundi RS et al., Varma S et al., Ankur Singh et al., R RS et al., Kansara AH et al., Kumar KM et al., Kausar Z et al. [6,7,8,9,10,11,12].

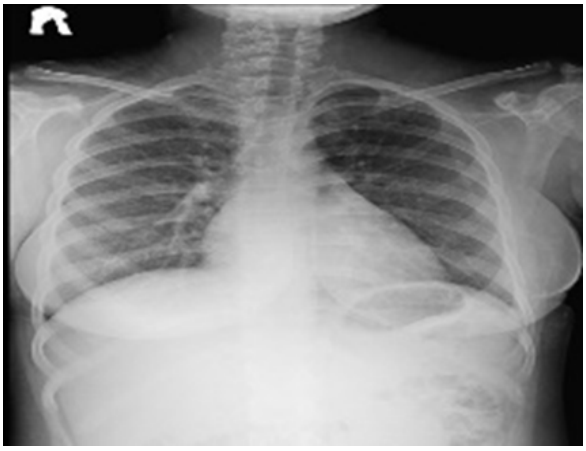
Surgical excision is the treatment modality of choice which was performed in almost every case including present study with better outcome [6,7,9,10,11,12]. Immunohistochemistry was done in present study but was not found to be done in other referred literature. This discrepancy may be due to limited financial support and or scarcity of resources.

CONCLUSION

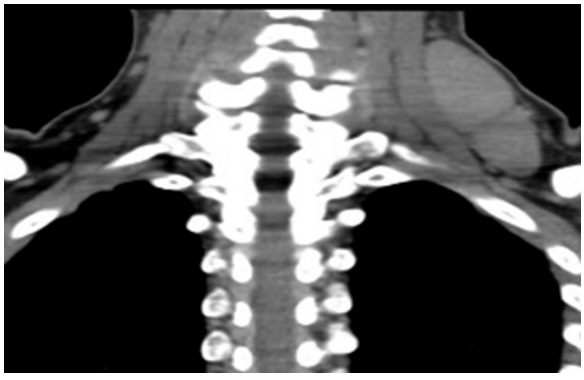
Unicentric hyaline vascular variant of castleman disease is often curable with surgery. Concerning the possibility of malignant transformation (lymphoma), close follow-up is mandatory. Overall presentation, histological type, treatment modality and prognosis among different age groups is similar.

Key Message

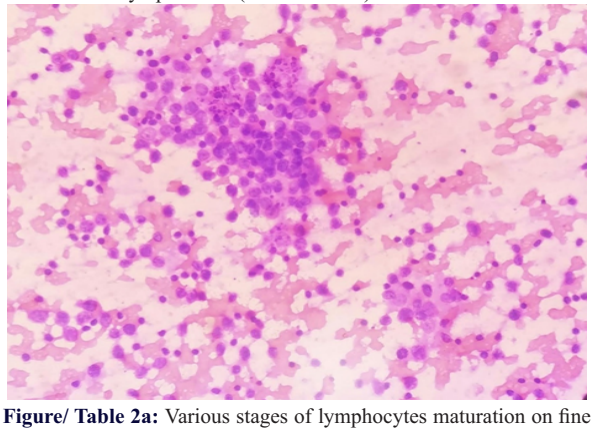
It should be considered in the differential diagnosis of lymphoma, metastatic adenopathy, and infectious and/ or inflammatory diseases that result in lymphadenopathy.



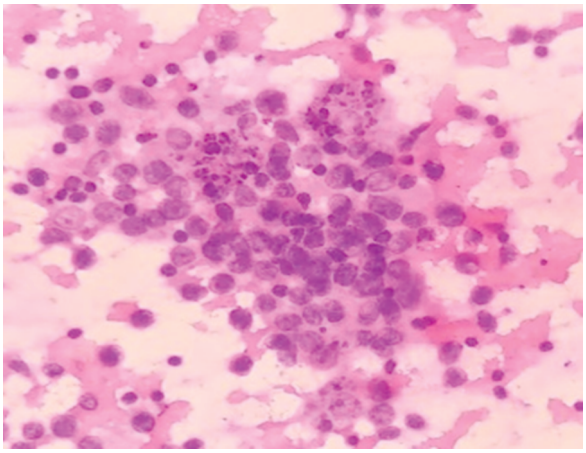
Figure/ Table 1a: Soft tissue swelling on left side of neck (CXR)



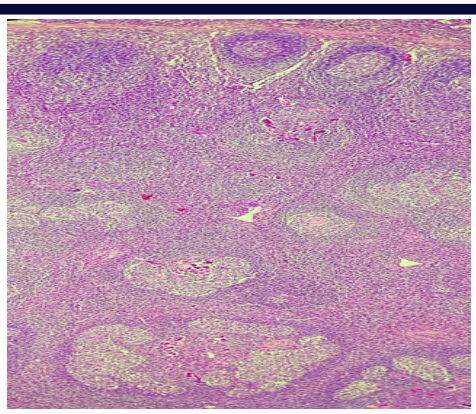
Figure/ Table 1b: Enlarged Well Defined Homogeneously Enhancing Left Cervical Lymph Nodes (CECT Thorax)



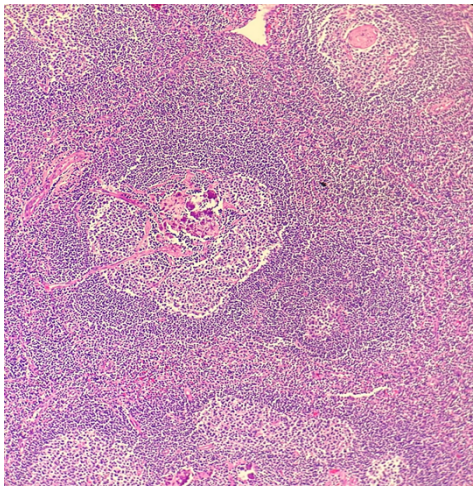
Figure/ Table 2a: Various stages of lymphocytes maturation on fine needle aspiration (10X, H&E)



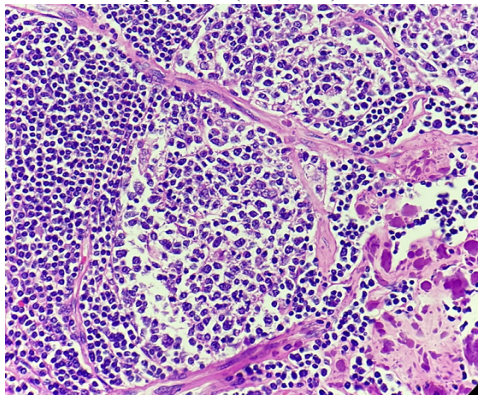
Figure/ Table 2b: Tingible body macrophages on fine needle aspiration (40X, H&E)



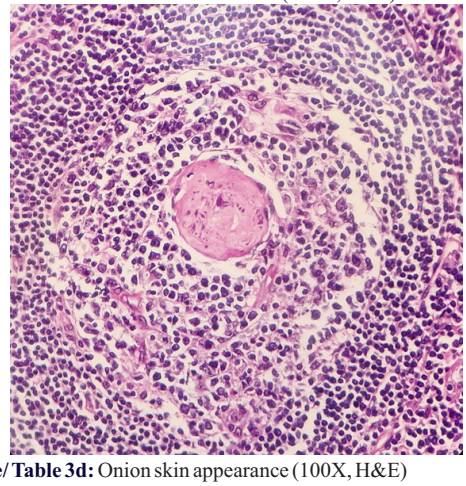
Figure/ Table 3a: Twinning (4X, H&E)



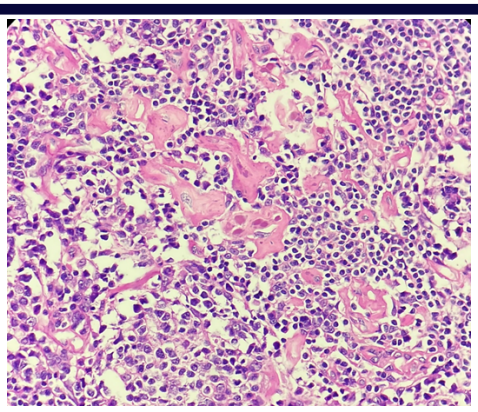
Figure/ Table 3b: Lollipop lesion (10X, H&E)



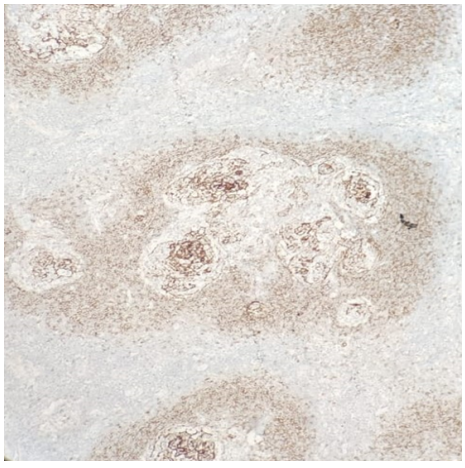
Figure/ Table 3c: Sclerotic blood vessel (100X, H&E)



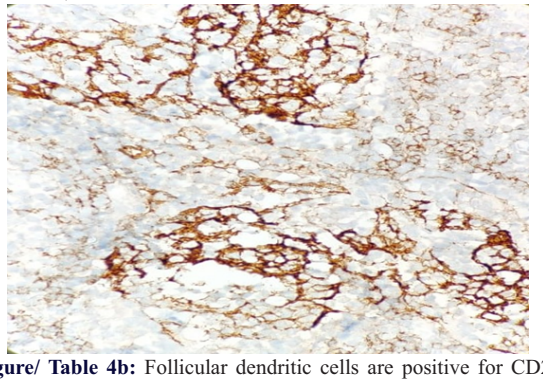
Figure/ Table 3d: Onion skin appearance (100X, H&E)



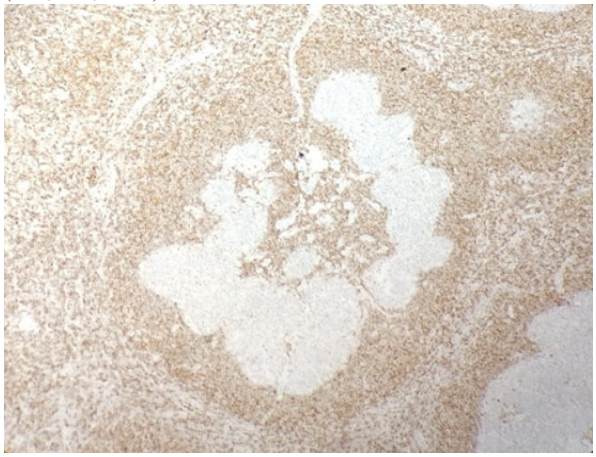
Figure/ Table 3e: Hyalinized stroma (40X, H&E)



Figure/ Table 4a: Follicular dendritic cells positive for CD23 (10X, IHC, Cd23)



Figure/ Table 4b: Follicular dendritic cells are positive for CD23 (40X, IHC, CD23)



Figure/ Table 4c: Germinal centres are negative for BCL-2 (10X, IHC, BCL-2)

Figure/ Table 5: Comparison of clinical presentation, histological type, treatment modality and outcome of Indian patients with present study.

Serial number	Parameter	Melkundi RS et al., [6]	Varma S et al., [7]	Ankur Singh et al., [8]	R RS et al.,[9]	Kansara AH et al., [10]	Kumar KM et al., [11]	Kausar Z et al., [12]	Present study
1	Age (in years)	18	14	04	12	12	08	13	16
2	Gender	Female	Female	Female	Male	Male	Male	Male	Female
3	Mode of presentation	Right sided neck mass	General ized seizures	-	-	-	-	-	Hole in heart with difficulty in breathing and difficulty in walking with discomfort in lower back
4	Enlarging neck mass	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
5	Location	Not specified	Cervical	Both sides of neck	Not specified	Anterior triangle of neck	Not specified	Not specified	Cervical level II
6	Size	4x2x5 cm	Not specified	4x3cm (left), 2x2 cm (right)	7x4 cm	4x6 cm	4x3.5 cm	3x1.5x1 cm	4x3x2cm
7	Site	Right	Right	Bilateral	Right	Left	Right	Right	Left
8	Most common imaging modality performed for neck nodes	CT Scan	Not done	Not done	Not done	Not done	USG	Not done	USG> CT scan
9	FNAC	moderate cellularity comprising of small lymphocytes in dys-cohesive and in small clusters	Not done	Polymorph ous	Reactive hyperplasi a	Not done	small and large lymphocytes, tingible body macrophages admixed with fibrotic strands in haemorrhagic background	Granuloma tous inflam mation	Reactive hyperplasia
10	Histopathology	Hyaline vascular	Hyaline vascular	population of	Hyaline vascular	Hyaline vascular	Hyaline vascular	Hyaline vascular	Hyaline vascular
11	IHC	Not mentioned	Not mentioned	lymphoid cells	Not mentioned	Not mentioned	Not mentioned	Not mentioned	Reactive in nature
12	Treatment modality	Excision	Only for seizures	Hyaline vascular	Excision	Excision	Excision	Excision	Excision
13	Prognosis with follow up	No Recurrence found after 1 year follow up	Not mentioned	Not mentioned	6 months after surgery, asympto matic	Not mentioned	6 months follow up; no recurrence	Not mentioned	No recurrence till present moment

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