Case Study

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Lymphadenopathy Presenting As Rosai- Dorfman- Destombes Disease; A Rare Entity

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1. Abstract

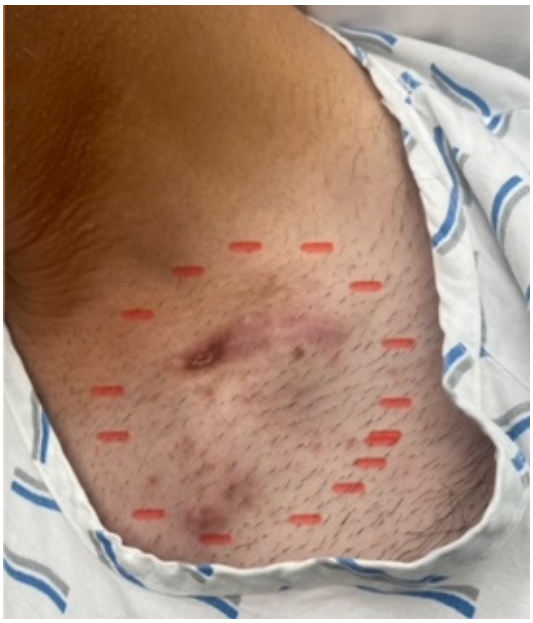
Rosai-Dorfman-Destombes disease is a very rare benign disorder of histiocyte proliferation with unknown etiology. We report the case of a 41 years old man diagnose with this disease, having had long standing cervical lymphadenopathy increasing in size. This case is presented given the difficulty in diagnosis and surgical management on skin involvement [1].

2. Introduction

Rosai-Dorfman- Destombes disease has a predilection of affecting cervical lymph nodes in adolescents and young adult patients but can have extra-nodal manifestation in 40-45% of cases. Although is a benign disease, and can be observed in up to 30% of cases, its rarity and difficulty of diagnosing adds up to the difficulty of management when needed. In view of mitigating such difficulties a consensus of diagnosis and management was established in 2018 under the tutelage of Histiocyte Society Meeting International Group. Our case reflects the importance of multidisciplinary team approach.

3. Case report

41 years old man, otherwise fit, relates a left supraclavicular swelling noticed 6 months back. No history of trauma. It was not painful but it was increasing in size and was associated with intermittent fever, no weakness described. Incidental notice of increase in size of the swelling after starting gym exercise. There was no restriction in the movement of the left shoulder. No past medical or surgical history. No history of recent travel. Clinical examination showed a left supraclavicular mass of 3 by 3 cm fixed in the subjacent tissues with associated skin discoloration.



Investigation:

4. MRI showed

Ill-defined soft tissue lesion measuring 3.3 x 3.4 cm noted in the left supraclavicular area surrounding the mid 1/3 of the underlying clavicle with no signs of invasion could be detected, seen inseparable from the upper aspect of the left pectoralis major muscle. The mass appears as isointense to the muscle in T1, slightly hyperintense in T2 and shows heterogeneous contrast up take.

Hepatitis B Surface Antigen	**NON REACTIVE**		
NON REACTIVE			
HIV 1 & 2 Ag & Ab	NON REACTIVE		
NON REACTIVE			
Hepatitis C Antibodies	NON REACTIVE		
NON REACTIVE			
WBC COUNT	6.9	6.9	9.9

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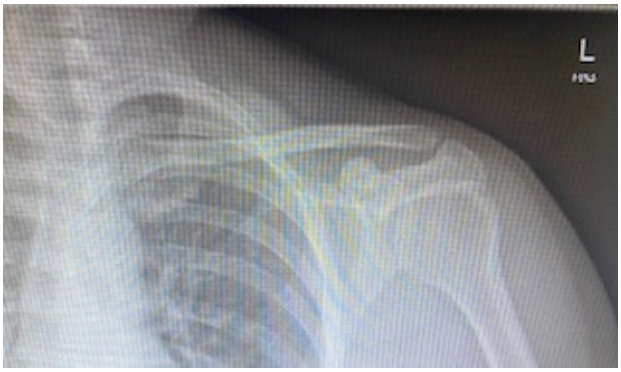
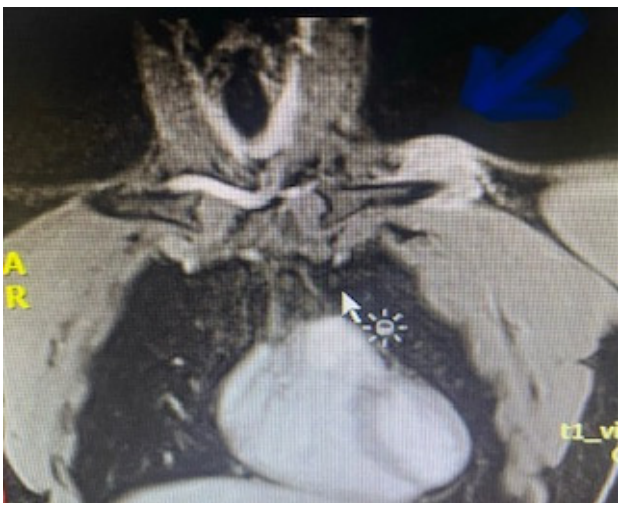
3.6 - 11.0 10^3/uL			
RBC COUNT	5.09	5.28 R	4.69 R
4.50 - 5.50 10^6/uL			
H E M O G L O B I N , BLOOD	13.3	14.3 R	12.2 Panic R
13.0 - 17.0 g/dL			
HEMATOCRIT	40.9	43.9 R	37.3 Panic R
40.0 - 50.0 %			
MCV	80.4	83.2 R	79.6 R, CM
77.0 - 95.0 fL			
MCH	26.2 Low	27.2 R	26.1 R, CM
27.0 - 32.0 pg			
MCHC	32.6	32.7 R	32.8 R, CM
31.5 - 34.5 g/dL			
RDW	13.7	12.7	12.8
11.5 - 14.0 %			
PLATELETS COUNT	396	278 R	252 R
150 - 410 10^3/uL			
MPV	7.5	8	7.7 CM
7.4 - 10.4 fL			
NEUTROPHIL Absolute	4.1	4.2 R	7.6 R, CM
2.0 - 7.0 10^3/uL			
Lymphocytes Absolute	2.1	2.1 R	1.4 R
1.0 - 3.0 10^3/uL			
Monocytes Absolute	0.5	0.4 R	0.9 R
0.2 - 1.0 10^3/uL			
Eosinophils Absolute	0.1	0.1	0
0.0 - 0.5 10^3/uL			
Basophils Absolute	0.1	0.1 R	0.0 Panic R
0.0 - 0.1 10^3/uL			
NEUTROPHIL%	59.6	60.8 R	76.6 Panic R, CM
%			
LYMPHOCYTE %	30.4	30.6 R	13.7 Panic R, CM
%			
MONOCYTE %	7.4	6.3 R	9.1 R
%			
EOSINOPHIL %	1.4	1.5 R	0.5 Panic R
%			
BASOPHIL %	1.2	0.8Panic R	0.1 Panic R
%			
Differential Type	AUTO DIFF	CM	
Nucleated RBC'S	0		
0 Per100WBC			

INR	0.95		
0.80 - 1.20			
PT Control	13.2		
APTT	45 High		
28.6 - 38.2 Secs			
Anti-Nuclear Antibody (ANA) by IFA	NEGATIVE < 1/100		
Cytomegalovirus IgG	POSITIVE Abnormal		
NEGATIVE			
Epstein Barr Virus DNA PCR - Blood	Detected (Positive		
Not detected (Negative)			

Diffusion restriction is noted. Enlarged left supra clavicular lymph node measuring 12 mm in diameter is seen. Focal area of bright marrow signal noted in the left humoral head? Bone cyst. (f 3,4,5) the initial core biopsy under ultrasound guidance was not specific with cores of fibrofatty, skeletal muscle and granulation tissue, no granulomas, but presence of Immunohistochemistry study (CKAE1/AE3, LCA and CD68): Negative for carcinoma Patient underwent incisional biopsy for diagnostic purpose with encounter of technical difficulties due to location as lump was encasing major blood vessels in the neck and inferiorly towards the apex of the lung.





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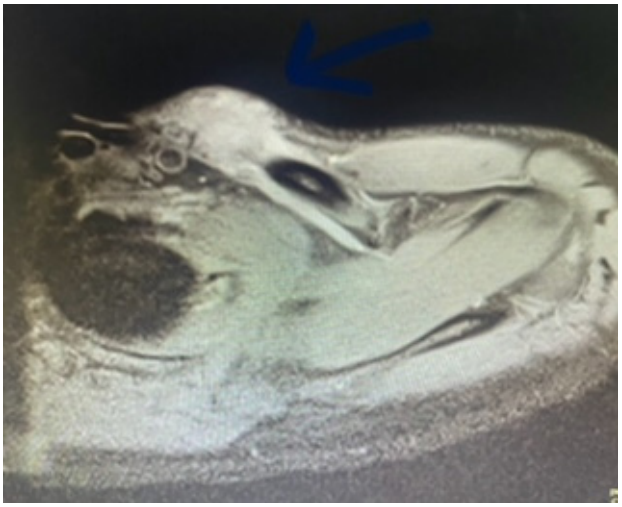

USS abdomen
Chest X ray:
Soft tissue swelling is seen projected over the middle third of left clavicle.
- Intact joint space and articulating margins of the left gleno-humeral and acromio-clavicular joints with no abnormalities detected.
- No periarticular soft tissue calcification could be seen.
- No fracture line or dislocation could be identified.

Clinical Details left supra clavicle lesion excisional biopsy was done Nature and Site of Specimen(s) Soft Tissue Gross Description Labelled with patient details and left supraclavicular lesion: Received in formalin are three fragments of irregular hemorrhagic tissue the largest 2.0 x 1.5 x 1.0 cm and smallest measuring 1.0 cm in diameter. No orientation. No skin is associated with this specimen. A1= Largest fragments. A2= other two fragments. 2 cassettes (all embedded). Grossed by Dr. Zuhair Elfadil. Microscopic Description The specimen was processed in its entirety in 2 blocks each examined at one level. Sections show lymphohistiocytic infiltrates with plasma cells and granulocytes in a background of sclerotic stroma. Zonation with pale and dark areas can be appreciated at low power examination. The darker areas are rich in small lymphocytes and plasma cells while the pale areas are rich in histiocytes with abundant vacuolated cytoplasm. Large histiocytes engulfing leucocytes are occasionally present (emperipolesis). Rare focal microabscess formation is also seen. Native lymph node structures are not seen; therefore a lymph node origin of the specimen cannot be confirmed. Granulomas are not identified and there is no evidence of metastatic carcinoma (AE 1/3 M immunohistochemistry stain performed) or metastatic germ cell tumour (OCT 3/4 immunohistochemistry stain performed). The large foamy histiocytes are positive for S-100, CD68 and OCT-2. They are negative for CD1a. The lymphoid cells are a mix of T and B lymphocytes (CD3, CD5, CD20, CD79a immunohistochemistry stains performed). Very occasional reactive lymphoid follicles are seen with germinal centers highlighted on CD21 immunohistochemistry stained section [2]. Ki 67 proliferation index is low. CD30 shows no evidence of RS cells and CD-15 stains scattered granulocytes only. PAS is negative for fungal hyphae, Giemsa stain is negative for parasites and Von Kossa stained section shows no evidence of Michaelis-Gutmann bodies to suggest Malakoplakia. Conclusion Left supraclavicular lesion biopsy: Rosai-Dorfman disease, see comments. Comment - The overall morphology is in keeping with Rosai-Dorfman disease, formerly known as (sinus histiocytosis with massive lymphadenopathy). - I favour extranodal over nodal disease. - This case was also seen by Dr Badr Hamid who agrees on the above diagnosis. - Please visit this link for some further information: The histopathology confirmed Rosai-Dorfman disease, formerly known as (sinus histiocytosis with massive lymphadenopathy). In favor of extra nodal rather than nodal disease. (F6) Patient underwent full excision of the lump with local plastic reconstruction of the skin and subcutaneous layers. At present on regular follow up.

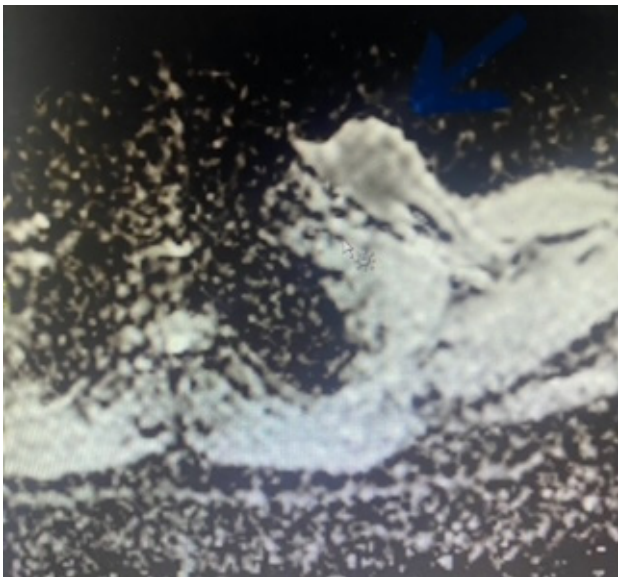

Figure 6:

Impression (conclusion or diagnosis)
Soft tissue swelling projected over the left clavicle for correlation with targeted soft tissue ultrasound

5. Discussion

Rosai -Dorfman- Destombes disease was first described [3] by Destombes in 1960s, who, as a pathologist, names it 'adenitis with lipid excess'. At present there are just above 1000 cases described in the literature[4]. Is a self-limited benign disease of unknown etiology? Under revised histiocytosis classification on 2016 is an R group disease of non-Langerhans histiocytosis. Clinically can occur as an isolated disorder or in association with autoimmune, hereditary, and malignant diseases. Literature [5] is citing prevalence of 1:200 000, is more frequently seen in children and young adults, although it has been reported up to age 74 years. Our case is a male of 41 years of age. Literature [6] emphasizes adequate tissue sample with possibility of flow cytometry and cytogenetic testing. There are cases reported based on FNAC[7]. Our case did not yield an appropriate diagnosis on core biopsy. Therefore, the specific

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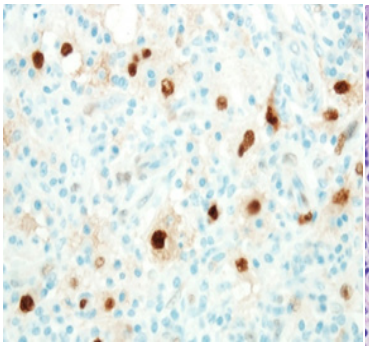
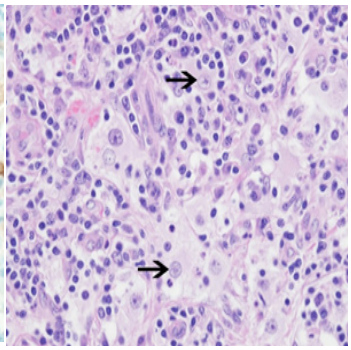
histopathology results of ample pale or "watery-clear" cytoplasm with a large hypo chromatic nucleus and prominent nucleolus was established on incisional biopsy with adequate sample.

The management [8] of this disease ranges from observation to extensive surgery or various regimens of medication- whether immunosuppressants, or chemotherapeutic agents. Overall, treatment is tailored on case to case basis. Observation is the rule, as 20-50% of the cases have spontaneous regression, especially on the nodal/cutaneous disease. This approach is particularly fit for the minimally symptomatic cases or uncomplicated nodal disease. The multidisciplinary [9] team work is essential not only for evaluation but for the management. Our case was considered initially for observation. Since patient was symptomatic on the area, he underwent full excision with skin reconstruction. In his case the option was considered with curative intent. The position of the mass, and particularities of the gross encasement of left supraclavicular vessels, posed difficulty in the technique, requiring thoracic, vascular and plastic surgery team in another facility, abroad.



*(S100 immuno-histochemistry. The inflammatory Histocytes are positive for S100)



*(Large histocytes engulfing cells -emperipolesis)

6. Conclusion

Rosai -Dorfman- Detumbles disease is a rare encounter with ongoing international Registry for Rare Histiocytic Disorders. It needs multidisciplinary team approach with careful medical history, examination, imaging, lab evaluation as it poses many challenges diagnostic wise, as well as therapeutic. While many cases can be observed, patients may require various treatment modalities, surgery being considered often with curative intent especially in the nodal and subcutaneous manifestations. The possible association of RDD with other immunological, hematological or oncological diseases further complicates its management. At present, the genetic mapping and mutational landscape of the disease is under investigation along with various promising targeted therapies.

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