TRIDHA
Journal of Clinical Cases & Reports
Volume 2022, Issue S6, 5 pages
https://doi.org/10.46619/joccr.2022.5-S6.1036

CASE REPORT

Kimura's Disease (KD) is an Unusual Cause of Head and Neck Masses

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Received: 20 April 2022; Accepted: 07 May 2022; Published: 14 May 2022

ABSTRACT

A 35-years-old male patient presented with unilateral fascial swelling for 4 years. On examination, smooth, firm mass not associated with thrill nor neurological deficits. Peripheral blood examination: peripheral eosinophilia. Acute phase reactant; normal, elevated serum Ig E level. Superficial US on neck; area of subcutaneous oedema involving the right check, reactive cervical lymph node. MRI with contrast; inflammatory pseudotumor.

Histopathological examination; Angiolymphoid hyperplasia with eosinophilia, no atypia nor malignant cell. Patient has (Peripheral eosinophilia, Elevated IgE and Biopsy; Angiolymphoid hyperplasia) diagnosed as Kimura's Disease and started steroid and cyclosporine with dramatic decrease in size.

INTRODUCTION

Kimura's Disease (KD) is a rare chronic inflammatory disorder presenting as multiple painless solitary subcutaneous nodules, predominantly in the head and neck region. KD characterized by painless nature and indolent course [1]. Radiologically KD may mimic other chronic inflammatory conditions and neoplasms, so histological evaluation along with elevated peripheral eosinophil and serum Ig E level are important for confirmatory diagnosis [2].

CASE REPORT

A 35-years-old male patient, married with two offspring, with no special habits of medical importance. No past history of diabetes, hypertension or chronic disease of medical importance with no history of drug addiction. Presented with unilateral fascial swelling for 4 years, steadily increase in size lead to fascial asymmetry. Patient reported that mass increase in size with any inflammatory condition e.g., common cold and disappeared after taking steroid and anti-inflammatory medication but recurrent again several times over years.

On examination, asymmetrical face with smooth, firm dome shaped mass in the right cheek, about (5 cm × 7 cm), normal overlying skin, not associated with thrill nor neurological deficits and multiple bilateral enlarged cervical lymph node with no abnormality detected in abdomen, heart, CNS and chest examination.

Peripheral blood examination: normal WBCs, HB, Platelets except peripheral eosinophilia. Acute phase reactant; normal (ESR $51^{st}/162^{nd}$), (CRP = 48, positive) elevated serum Ig E level (200 IU/ml), Ig G normal. Serology (ANA, RF, AntidsDNA, ANCA-C, ANCA-P) was normal.

Superficial US on neck; area of subcutaneous oedema (4.9 cm × 2.5 cm) involving the right check with few enlarged reactive deep cervical lymph nodes with preserved shape and hilum. MRI with contrast; ill-defined soft tissue at right side of check reaching downwards the right submandibular region with heterogenous contrast enhancement; inflammatory pseudotumor and multiple enlarged submental, right submandibular, right intra-parotid and bilateral deep cervical benign lymph nodes.

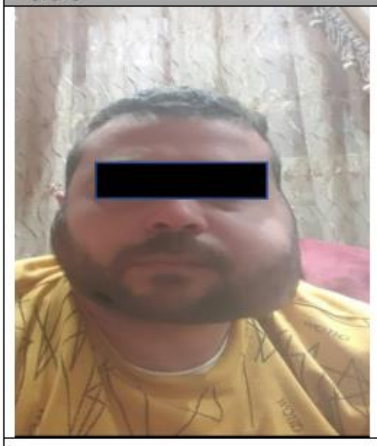
Histopathological examination; Angiolymphoid hyperplasia with eosinophilia infiltration, no atypia nor malignant cell.

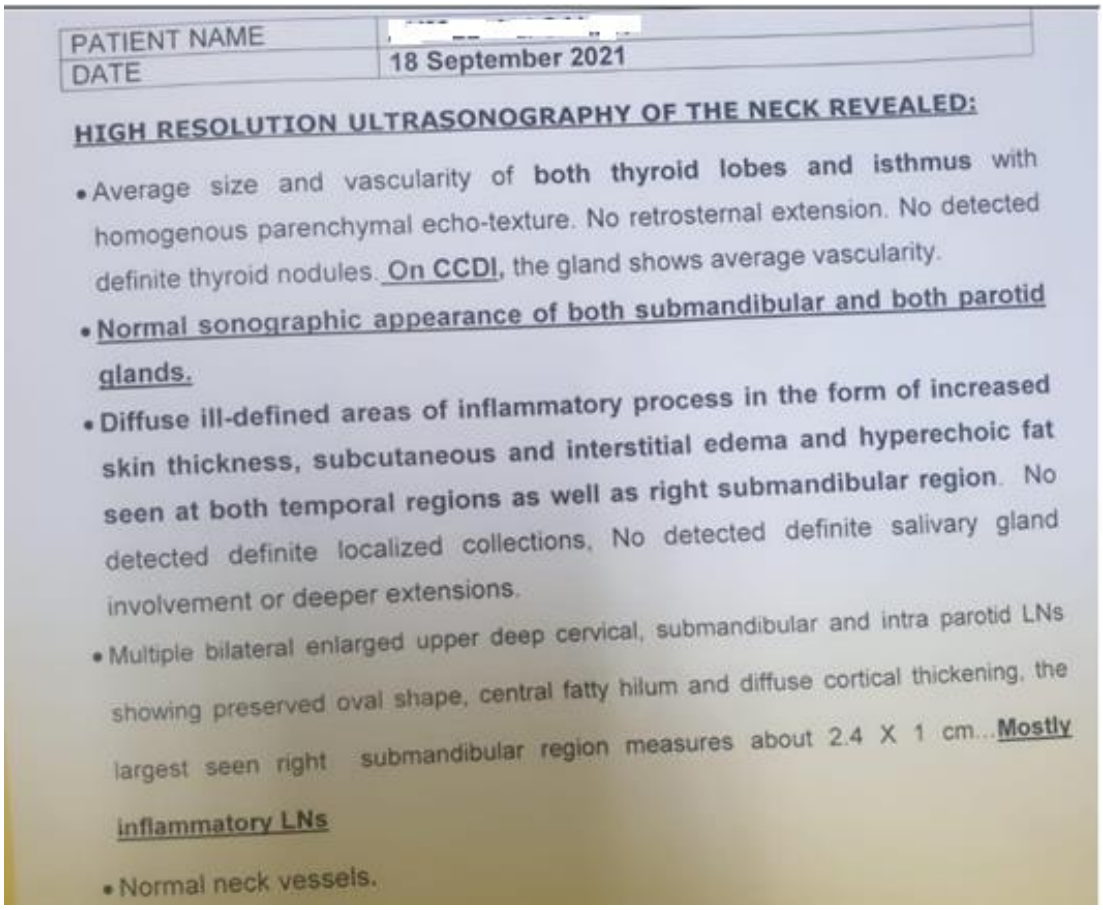
In our case, Patient has (Peripheral eosinophilia, Elevated Ig E and Biopsy; Angiolymphoid hyperplasia), after exclusion of (Oral and Odontogenic infection, Lipoma, Inflammatory lesion of salivary gland, IgG4 Related disease, Sarcoidosis, Sjogren syndrome, Hodgkin and Non-Hodgkin lymphoma) patient diagnosed as Kimura's disease and started steroid and cyclosporine with dramatic decrease in size, complete remission over few months.

DISCUSSION

Kimura's disease (KD) is an unusual vascular tumor that typically occurs at 20 years to 30 years of age [3] characterized by a triad of painless subcutaneous masses in the head or neck region, blood and tissue eosinophilia, and markedly elevated serum immunoglobulin E levels. The presentation may be associated with renal disease in 20% patients [4], nephrotic syndrome. Definitive diagnosis of KD is based on histological findings [5] because imaging studies mimicking vascular malformation and Hodgkin's lymphoma. The optimal treatment of KD is controversial due to lack of large-scale systemic clinical studies, Successful cases have been reported by applying surgery with 46.2% recurrent risk, radiotherapy (used in recurrent and persistent cases) and chemotherapy. Medical therapy, cyclosporine effective to induce remission, Corticosteroid are commonly used but with high risk for recurrence after cessation. IVIG as potent steroid sparing agent, maintain disease free more than 6 years.

Our patient diagnosed as Kimura's disease based on Peripheral eosinophilia, Elevated IgE and Angiolymphoid hyperplasia on histological examination [6] with good response to cyclosporine and corticosteroid.

Before	After
	
Figure(1): The face appeared asymmetrical with a swelling in the right cheek which was large in size about (5*7cm),smooth ,dome shaped **Permission was taken from patient for publication**	Figure (2): as comprasion to before, significant decrease in the size of mass,with more or less symmetrical face.



PATIENT NAME [redacted]

DATE 18 September 2021

HIGH RESOLUTION ULTRASONOGRAPHY OF THE NECK REVEALED:

- Average size and vascularity of **both thyroid lobes and isthmus** with homogenous parenchymal echo-texture. No retrosternal extension. No detected definite thyroid nodules. **On CCDI**, the gland shows average vascularity.
- **Normal sonographic appearance of both submandibular and both parotid glands.**
- **Diffuse ill-defined areas of inflammatory process in the form of increased skin thickness, subcutaneous and interstitial edema and hyperechoic fat seen at both temporal regions as well as right submandibular region**. No detected definite localized collections. No detected definite salivary gland involvement or deeper extensions.
- Multiple bilateral enlarged upper deep cervical, submandibular and intra parotid LNs showing preserved oval shape, central fatty hilum and diffuse cortical thickening, the largest seen right submandibular region measures about 2.4 X 1 cm..**Mostly inflammatory LNs**
- Normal neck vessels.

Figure 3: Neck US; inflammatory mass at both temporaland right submandibular region.

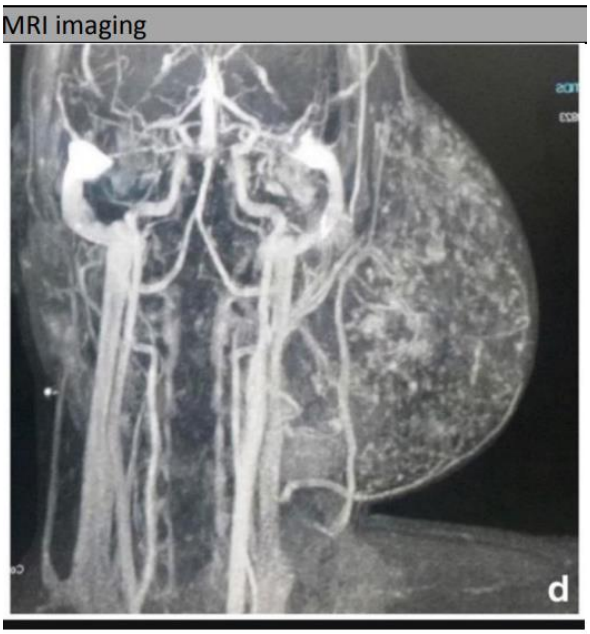


Figure 4: MRI imaging of the mass show inflammatory mass.

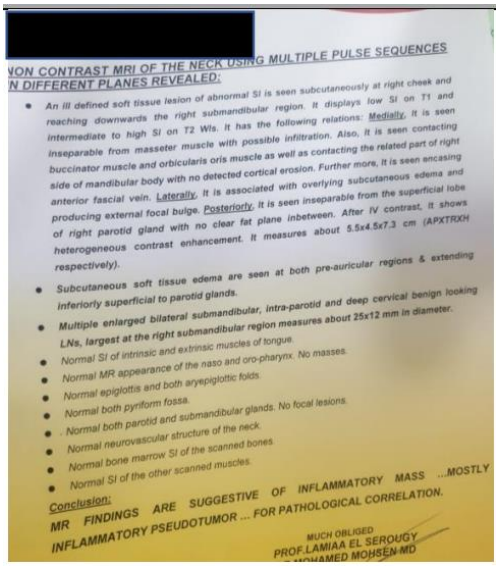
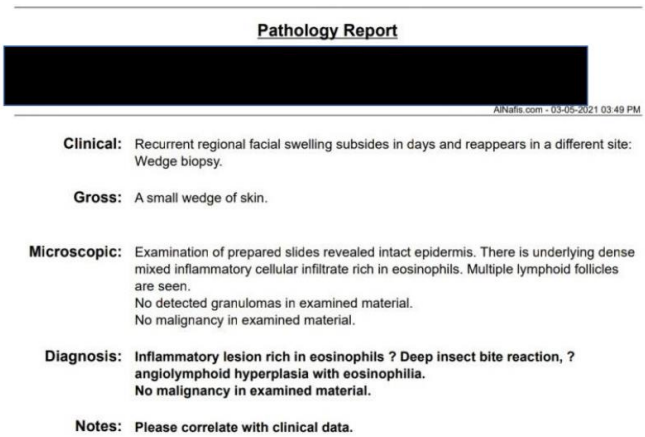


Figure 5: Non contrast MRI show inflammatory pseudotumor.



Pathology Report

[redacted]

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Clinical: Recurrent regional facial swelling subsides in days and reappears in a different site: Wedge biopsy.

Gross: A small wedge of skin.

Microscopic: Examination of prepared slides revealed intact epidermis. There is underlying dense mixed inflammatory cellular infiltrate rich in eosinophils. Multiple lymphoid follicles are seen.
No detected granulomas in examined material.
No malignancy in examined material.

Diagnosis: **Inflammatory lesion rich in eosinophils ? Deep insect bite reaction, ? angiolymphoid hyperplasia with eosinophilia.**
No malignancy in examined material.

Notes: **Please correlate with clinical data.**

Figure 6: Pathological report shows angiolymphoid hyperplasia with eosinophilia.

CONCLUSION

Patients with subcutaneous painless soft tissue swelling in the head and neck, a high possibility of KD considered if peripheral blood eosinophils increase, especially when IgE levels increase in the serum.

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