Castelman disease: an unusual presentation of a rare disease

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INTRODUCTION

- Castleman's disease is a rare group of lymphoproliferative disorders characterized by angiofollicular lymph node hypertrophy.
- Unicentric CD can be cured by excision surgery or radiotherapy and can be challenging in diagnosis.

AIM

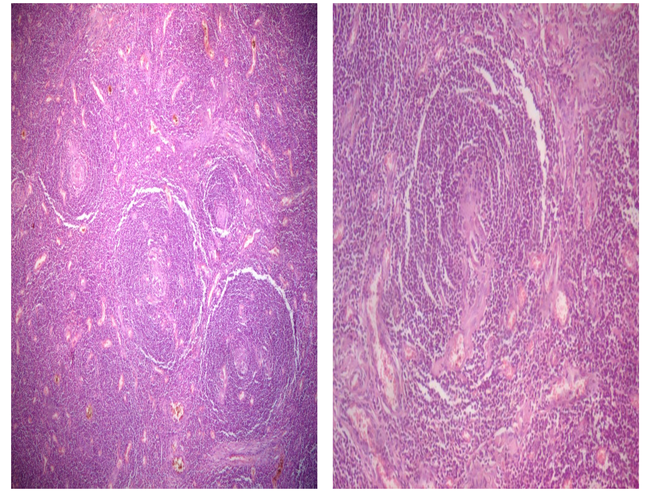
we report the clinicopathologic findings and treatment outcome of this unusual presentation of an Egyptian young gentleman who was diagnosed UCD and his treatment journey.

METHOD

- A case report which includes diagnostic history taking, physical examination, full blood count, chemistry profile, virology, immune profile, tumor markers and tissue biopsy.
- This came along with radiological follow up.

RESULTS

- Our 28 year old gentleman presented with a one year history of fever and abdominal pain.
- Ultrasound scan showed gastric wall mass sized 5*6 cm arising from inner aspect of posterior wall of pyloric region 4*6 cm, highly vascular at periphery with nodal infiltration which was highly suspicious of gastrointestinal stromal tumor (GIST).
- Hence, Laparoscopic total excisional biopsy finally diagnosed it as Castelman disease (hyaline vascular type) with reactive hyperplasia of associated LNs. Autoimmune disorders, overlapping IgG4-related disease, viral infection; TAFRO syndrome and other malignancies were excluded.
- Our patient was classified as Unicentric Castelman disease treated with total excision. Follow up scans showed complete resolution with no residual lesions. He is now on an uneventful follow up at our outpatient clinic.



Photomicrograph (1): Atretic germinal centers surrounded by concentrically arranged lymphocyte in mantle zone (onion-skin appearance) traversed by sclerotic blood vessels with vascular proliferation in the interfollicular region (H&E; x100).

Photomicrograph (2): Hyalnized blood vessels penetrate the regressively transformed germinal centers at right angle giving the lollipop appearance (H&E; x100).

CONCLUSIONS

- UCD sometimes can be very challenging to diagnose and can be mis-diagnosed as solid malignancy especially on rare atypical presentations.
- The curative role of surgery in UCD has dramatically improved the outcome of this rare cohort of patients.

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