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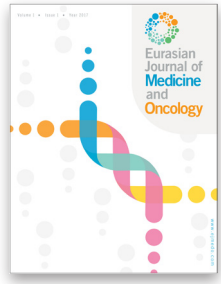

Case Report

Castleman's Disease Associated with Pemphigus Vulgaris: Remains Retrospectively Diagnosed Entity

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Abstract

Castleman's disease is a rare, benign, lymphoproliferative disorder of unknown cause.[1] The disease can be occasionally associated with a paraneoplastic pemphigus (PNP), an autoimmune mucocutaneous disorder commonly seen in neoplasms of lymphocytic origin.[2] Castleman's tumor has been found in only 10 % of PNP patients,[3] so the diagnosis depends mainly on the pathological examination. For pathological examination, complete surgical excision biopsy of uni-centric disease remains best mode of treatment. We report a case of retrospectively diagnosed case of castleman's disease with PNP after pathological examination.

Keywords: Castleman's disease, paraneoplastic pemphigus (PNP), histopathological examination.

Paraneoplastic pemphigus (PNP) is an autoimmune mucocutaneous disease associated with lymphoproliferative neoplasms, and frequently with a very rare tumour, Castleman's disease. There is enough literature now available to suspect castleman's disease when PMP is found along with mediastinal or retroperitoneal mass, however due to rarity of disease, diagnosis (is revealed on final histopathological report) remains curtailed till excisional pathological report comes. We are in reporting one case of retroperitoneal mass with oral mucocutaneous lesions diagnosed as castleman's disease in a retrospective manner.

Case Report

A 24-year-old male patient was referred to our Surgical OPD with the diagnosis of a large retroperitoneal pelvic tumor for further treatment. Patient's symptoms had begun six month earlier with rapidly progressive diffuse cutaneous lesions all over the trunk and extremities as well as oral lesions with the form of erythematous plaques and superficial hyperceratosis.

The patient had been initially treated with local corticosteroids, but skin lesions had remained stable after initial response.

A subsequently performed contrast-enhanced multisliceCT demonstrated a highly vascularized retroperitoneal mass (9.8×7.6 cm) with significant contrast enhancement consistent with a sarcoma or paraganglioma. The tumor located in left Para aortic space between aorta, common iliac vessels and left ureter.

On physical examination besides the above mentioned diffuse erosive muscositis (Fig. 1) with lichenoid inflammation, no lymphadenopathy or organomegaly was noted. Preoperative evaluation including Complete Blood Count, Liver Function Test, Renal Function Test and Prothrombin Time were normal.

Following thorough preoperative fitness the patient was taken to laparotomy for resection of the tumor. Sigmoid colon and descending colon mobilized lateral to medial and anterior surface of tumor was exposed. Tumor was dissect-

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ed free from aorta and common iliac vessels. Left ureter was safe guarded and complete tumor excision was done (Fig. 2). While excision patient lost around 1.8 liter of blood, so he required 6 units of PCV intraoperatively.

Patient was kept in ICU for two postoperative days. He had uneventful recovery in post-operative period and discharged on post-operative day seven. In consultation with Dermatologist, he was kept on oral steroids. Final histopathology report diagnosed hyaline variety of castleman's disease (Fig. 3).

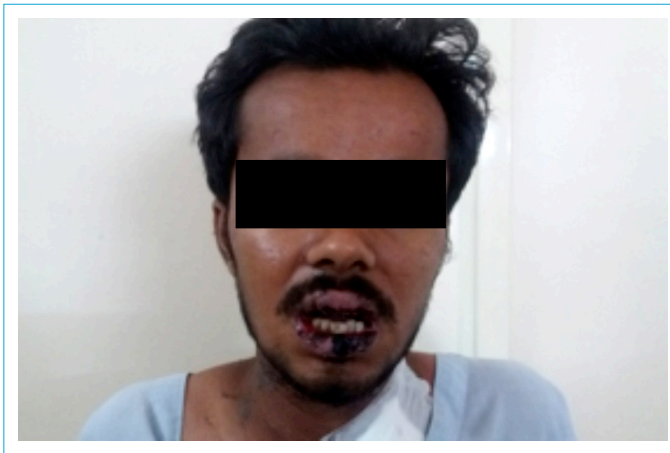

Figure 1. Mucocutaneous lesions.

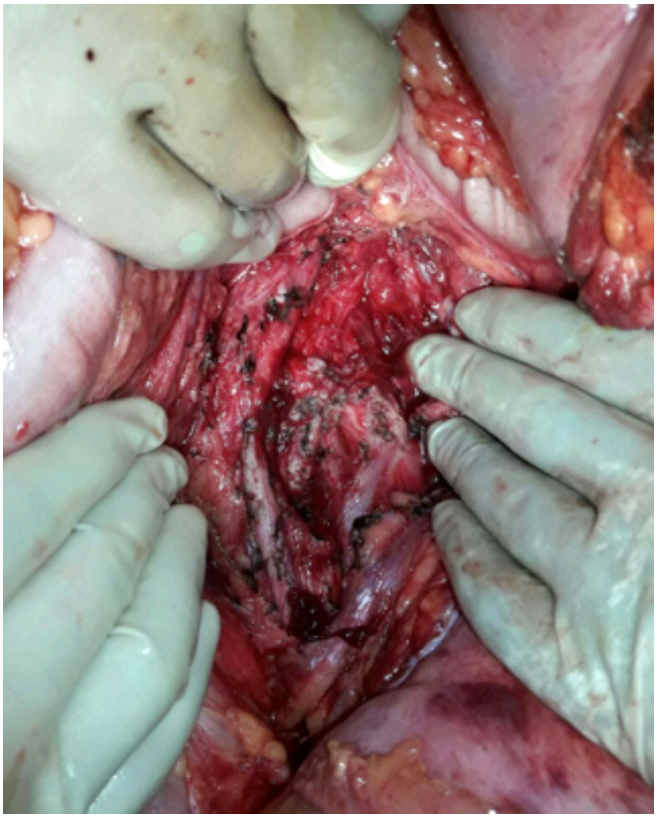

Figure 2. Intraoperative picture after excision.

Patient was readmitted after a week due to fresh eruptions of skin all over body. He was managed using intravenous steroids. He improved within 72 hours. He was discharged after 7 days on tapering regime of steroid. At follow up of one month, he was symptom free from cutaneous lesions.

Discussion

Castleman's tumors are neoplasms of lymphatic origin, also known as giant lymph node hyperplasia or benign giant lymphoma. There are three histological types of castleman's tumor: a) Hyaline-vascular type (80–90%), b) plasma cell type (10–20%), and c) intermediate types.[4] The most common location of the tumor is the mediastinum (60 – 70%). Abdominal forms are less common (10-20%). The majority abdominal location found at retroperitoneum. There are two variants of tumor: Localised and Multicentric. Multicentric variants have aggressive clinical course, systemic symptoms, organomegaly and neoplastic transformation have been reported. Castleman's disease has been associated with a very high incidence of autoimmune phenomena such as cytopenia, peripheral neuropathy, systemic lupus erythematosus, Sjögren's syndrome, and myasthenia gravis.[5] Our case was diagnosed as hyaline type of histological variant.

PNP is a clinically, histologically and immunologically distinct autoimmune mucocutaneous disease.[6] A variety of neoplasms have been reported in PNP, more commonly with haematological diseases including Non-Hodgkin Lymphoma, chronic lymphocytic leukaemia, and Waldenstroems's macroglobulinaemia. PNP is rarely associated with solid tumors such as sarcoma, bronchial carcinoma, colonic dysplasia, Castleman's disease and thymoma.[7,8] Castleman's tumor

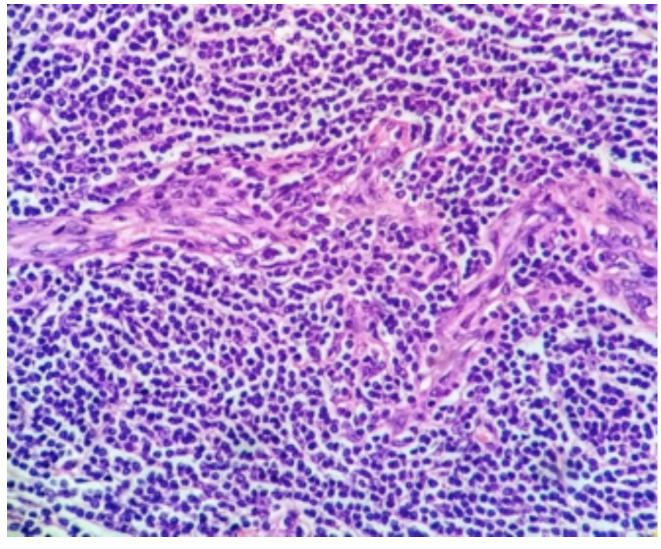

Figure 3. Hyaline variety of Castleman's disease.

has been found in approximately 10% of PNP patients.[4] The distinctive clinical findings in PNP include severe painful oral erosions and ulcerations with hemorrhagic crusting of the lips and polymorphous skin lesions resembling erythema multiforme, pemphigus vulgaris (PV) or lichen planus pemphigoides.[9] These clinical findings were observed in our patient. Cases of PNP with pulmonary involvement resulting in respiratory failure have been also reported.[9]

In our case the diagnosis of paraneoplastic pemphigus was retrospective. Initially it was described as lichen planus in association with pemphigus. In a review of literature, more than half cases diagnosed retrospectively.

Treatment of the disease depends mainly on the histological type and the clinical symptoms. Surgery is now the gold standard for the treatment of localized disease with curative results in most of the cases. High doses of corticosteroids, radiation, chemotherapy and immunosuppressive therapy have been used as additional therapeutic modalities. It should be however pointed out that PNP immunosuppressive treatment alone is ineffective

Without treatment of the underlying neoplasm. A complete remission of the skin lesions has been observed once the tumor has been removed obviating the need for further immunosuppressive treatment.[3, 7] This observation emphasizes on the role of surgeons in the treatment of the syndrome with an early and complete excision of the tumor in compliance with principles of surgical oncology. We have to point out that complete surgical resection, although curative, is in some cases precluded due to hypervascularity of the tumor or invasion of

adjacent structures.[1]

Conclusion

Castelman's disease should be first diffential diagnosis in patient with localised hypervascular tumor with pemphigus. Pemphigus not responding to medical treatment, an underlying tumor should be suspected and evaluated. Complete surgical resection of the localized tumor can be curative in most of the cases.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

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