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Case Report

Interesting features associated with Castleman disease: a case report

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Introduction

Castleman disease (CD) is a rare lymphoproliferative disorder. Multicentric CD is a systemic disease presenting with a multitude of symptoms. POEMS syndrome is a rare paraneoplastic syndrome due to an underlying plasma cell disorder. The acronym refers to the features of the syndrome: polyradiculoneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell disorder and skin changes [1]. Meningioma is a common neoplasm of the central nervous system [2] En-plaque meningioma can be a manifestation of Castleman disease [3]. We report a case of multicentric CD associated with POEMS syndrome and en-plaque meningioma.

Case report

A 68-year-old female with a history of long-standing hypertension presented with fever of one-week duration and abdominal distension.

On preliminary examination, she was afebrile and pale with coarse facies, finger clubbing, leukonychia, hypertrichosis over the limbs and bilateral ankle oedema. She had multiple cervical and axillary lymph nodes, which were 2cm × 3cm, firm, mobile and non-tender. There was bilateral mild pleural effusion. Abdominal examination revealed hepatosplenomegaly with moderate ascites. Neurologically she was found to have a high stepping gait, distal weakness of both legs (power of grade 4) with areflexia and glove and stocking type neuropathy. She was also found to have a right sided 6th nerve palsy without papilloedema. On further questioning, she revealed that she had diplopia, walking difficulty and loss of appetite for more than 6 months for which she did not seek any medical advice. Our working diagnosis was advanced lymphoma.

Blood investigations showed a haemoglobin of 9.8g/dL (12–16), fasting blood sugar of 102mg/dL, erythrocyte sedimentation rate (ESR) of 89mm, serum albumin of 33.4g/L and serum globulin of 38.3g/L. Ascitic fluid had a low serum/ ascites albumin gradient (ascitic fluid protein of 36.3g/L). Other basic blood investigations were normal. Tuberculosis screening by the Mantoux test, peritoneal fluid acid fast bacilli smear and culture and

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molecular testing by nucleic acid amplification via the GeneXpert system was negative. Retroviral serology was negative. Lymph node excision biopsy revealed Castleman disease of the hyaline vascular type. Immunohistochemistry was not done as there were no atypical cells in the lymph node excision biopsy.

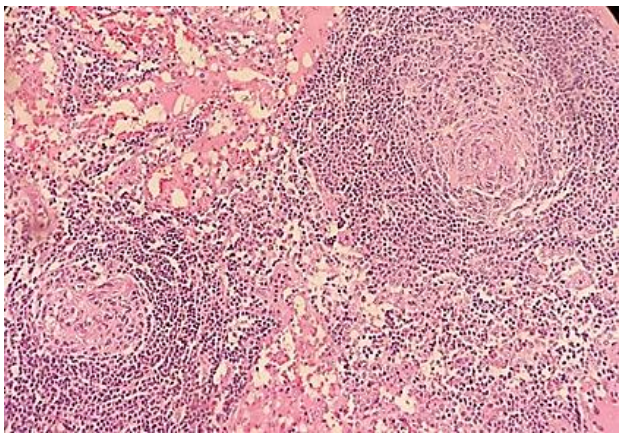

Figure 1: Histology of the lymph node shows tight concentric layering of the mantle cell layer with onion skin appearance. Germinal centers show hyalinization and vascular proliferation

After the tissue diagnosis, the clinical examination findings made us suspect POEMS syndrome. The immunoglobulin profile [IgG 2015mg/dL (569 -1919mg/dL), IgM 217mg/dL (47 – 147mg/dL) and IgA 151mg/dL (61 – 330mg/dL)] revealed dysimmunoglobulinemia. Serum protein electrophoresis showed monoclonal gammopathy. Bone marrow trephine biopsy showed 8% of plasma cells and megakaryopoiesis. Thyroid profile was suggestive of hypothyroidism but 9 am cortisol was normal. Nerve conduction studies demonstrated a demyelinating type of neuropathy. Contrast enhanced computerized tomography studies supported the clinical examination findings but did not reveal any masses. Echocardiography was normal apart from a thin rim of pericardial effusion. Magnetic resonance imaging (MRI) of the brain showed a right side sphenoid wing en-plaque meningioma with local invasion.

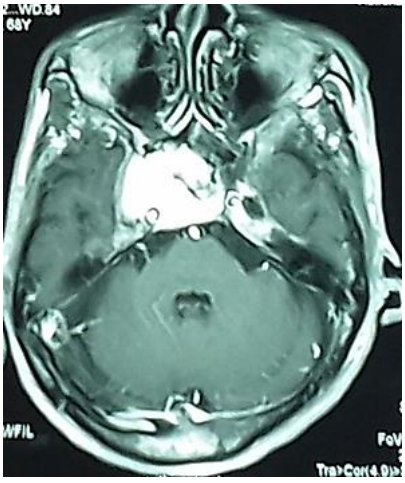

Figure 2: FLAIR contrast MRI; intermediate, homogenously contrast enhancing mass lesion arising from the right sphenoid wing with local infiltration**.**

Patient did not consent for a trans-sphenoidal biopsy of the en-plaque meningioma mass. She was referred to an oncologist and planned for radiotherapy but she refused treatment.

Discussion

CD is a rare disorder and occurs in 11 - 30% of patients with POEMS syndrome [4]. It involves non-malignant proliferation of the lymph nodes and affects patients of varying ages, typically 50 – 65 years [5]. There are two pathological types of CD, the hyaline vascular and the plasma cell variant [5]. The hyaline vascular type is the most common (80-90%). The plasma cell variant is seen in 10-20% of cases and a small percentage present with a mixed histologic appearance [1, 5]. Multicentric CD may be associated with human herpesvirus 8 (HHV-8) infections. In contrast, unicentric CD is a localized lymphadenopathy without HHV-8 association [5]. HHV-8 serology was not performed in this patient due to financial constraints. She was negative for retroviral serology.

Table 1: Criteria for the diagnosis of POEMS syndrome.

Mandatory major criteria	Polyneuropathy (typically demyelinating type)
	Monoclonal plasma cell proliferative disorder
Other major criteria (one required)	Castleman disease
	Sclerotic bone lesions
	Elevated vascular endothelial growth factor
Minor criteria	Organomegaly (hepatomegaly, splenomegaly or lymphadenopathy)
	Extravascular volume overload (oedema, pleural effusion, ascites)
	Endocrinopathy (adrenal, thyroid, pituitary, pancreatic, gonadal)
	Skin changes (hypertrichosis, white nails, hyperpigmentation, acrocyanosis)
	Papilloedema
	Thrombocytosis, polycythemia
Other signs and symptoms	Clubbing, weight loss, diarrhoea, thrombotic diatheses, hyperhidrosis, pulmonary hypertension

POEMS syndrome is a type of paraneoplastic syndrome. The diagnosis of POEMS is based on clinical and laboratory findings and it may be missed if it is not considered in the differential diagnosis. Our patient fulfilled most of the above criteria. Clubbing and exudative ascitic fluid are seen in POEMS and the bone marrow in POEMS often shows megakaryopoiesis [1]. These findings were observed in our patient.

Interestingly, the patient was found to have an en-plaque meningioma of the sphenoid wing. The lymphoplasmacyte-rich histological type meningioma characteristically arises as an en-plaque meningioma. It usually invades the other structures and may be associated with hypergammaglobulinaemia and anaemia [2]. CD is one of the differential diagnoses of lymphoplasmacyte- rich meningioma [6]. In our patient we could not biopsy the meningioma but the radiological findings supported the diagnosis of en -plaque meningioma.

Conclusion

CD should be considered as a differential diagnosis in a patient with multiple lymphadenopathies. POEMS can be missed easily. A good clinical history, thorough physical examination and supportive laboratory findings will help the correct diagnosis. Even though meningioma is a common tumour, it can be a presentation of CD.

Consent

Written informed consent was obtained from the patient for publication of this case report.

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Authors' contributions

MI, BS, JR analysed the clinical scenario and arrived at the final diagnosis. JN contributed in the literature survey and prepared the first draft. MI and BS reviewed and approved the manuscript. All authors provided clinical care for the patient. All authors read and approved the final manuscript.

Availability of data and materials

All the data supporting our case report is contained within the manuscript.

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