· Case report ·

Acute onset of proptosis secondary to subcutaneous panniculitis-like T cell lymphoma

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Abstract

• AIM: To report a rare case of acute onset of proptosis secondary to subcutaneous panniculitis-like T cell lymphoma (SPTCL).

• METHODS: A case report.

• RESULTS: A 27-year-old Malay man presented with history of acute onset of proptosis in the left eye for 2 weeks. It was associated with history of prolonged high fever. He also developed multiple erythematous swelling on his body and both thighs during this period. On examination, visual acuity in both eyes was 6/6. The left eye was proptosed and chemotic. The extraocular movement was limited in all directions. The cornea and the anterior segment were normal. Fundoscopy showed normal optic disc and retina. The examination of the right eye was unremarkable. His body temperature was 40.0℃. There was presence of multiple erythematous subcutaneous lesions over the body mainly in the left axillary, right hypochondriac, both thighs and the suprapubic area. The inguinal lympnodes were also palpable. MRI of orbit and brain revealed generalized soft tissue swelling in the left extraconal and retro-orbital space suggesting of inflammatory reaction. The cavernous sinus and brain were normal. Skin biopsy from the erythematous lesion revealed as SPTCL. He was referred to the haematologist and was started on chemotherapy-CHOP regime. The patient responded well to the chemotherapy and the proptosis reduced.

• CONCLUSION: Proptosis secondary to SPTCL is very rare. This is a variant of a peripheral T cell lymphoma characterised by multiple subcutaneous nodules presented with proptosis and fever.

• KEYWORDS: subcutaneous panniculitis-like T cell lymphoma; benign panniculitis; proptosis
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INTRODUCTION

Subcutaneous panniculitis-like T cell lymphoma (SPTCL) is a very rare skin lymphoma. It is characterized by infiltration of the subcutaneous tissue by neoplastic cytotoxic T cells mimicking panniculitis[1]. It was first described by Gonzales *et al*[2] in 1991. Commonly, young adults are affected, with median age of 36 years. There is a female predominance with male to female ratio of 0.5[1]. Proptosis as the primary presentation of SPTCL is very rare. A case of SPTCL is most likely to present to the dermatologist. However, we would like to report a rare case of SPTCL presented as proptosis.

CASE REPORT

A 27-year-old Malay man presented with history of acute onset of proptosis in the left eye for 2 weeks. It was associated with history of prolonged high fever and chills, generalized malaise and myalgia. He also developed multiple erythematous swelling on his body and both thighs. Some of the swelling resolved spontaneously at the time of the presentation. There was no significant medical and surgical history.

On examination, visual acuity in both eyes was 6/6. The left eye was proptosed and chemotic (Figure 1). The extraocular movements were limited in all directions. The cornea and the anterior segment were normal. Fundoscopy showed normal optic disc and retina. The examination of the right eye was unremarkable. The physical examination revealed a physically healthy young male with a body temperature of 40.0℃, pulse of 88 bpm and blood pressure of 110/70mmHg. His heart sounds were normal, without any murmurs, rubs, or gallops, and the lungs were clear to auscultation bilaterally. There were multiple sizes of erythematous subcutaneous lesions ranging from 2cm×3cm up to 5cm×8cm noted over his body mainly in the left axillary, right hypochondriac and both thighs. The largest lesion was at the left thigh (Figure 2). The lesions were slightly tender on palpation. The inguinal lymphnodes were also palpable. The abdominal examination revealed normal bowel sounds with no hepatosplenomegaly.

The laboratory analysis included a complete blood cell count and picture and a complete metabolic panel was performed. The full blood picture revealed normal white and red blood cells with no presence of blast cell. ESR and C-reactive protein were raised Basic metabolic panel and thyroid

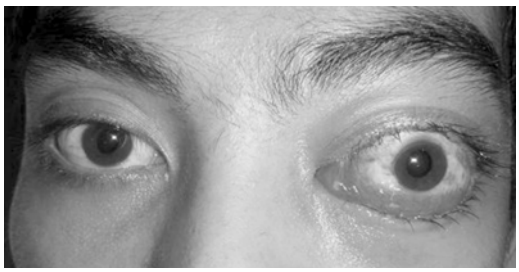

Figure 1 Patient presented with left eye chemosis and proptosis.

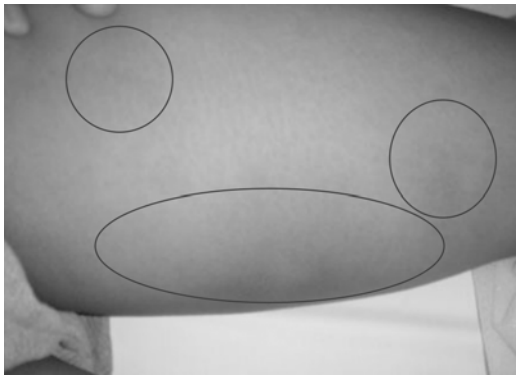

Figure 2 Erythematous subcutaneous lesions over the left thigh.

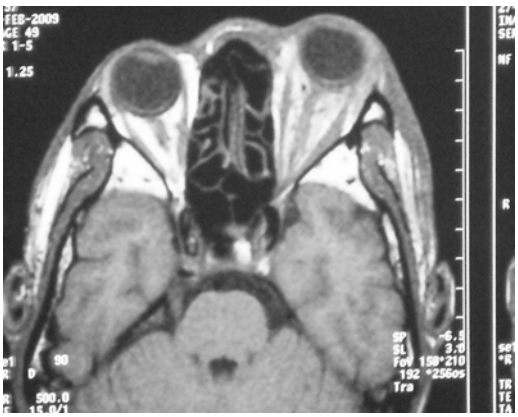

Figure 3 Axial view of MRI showing generalised soft tissue swelling in the left extraconal and retro-orbital space.

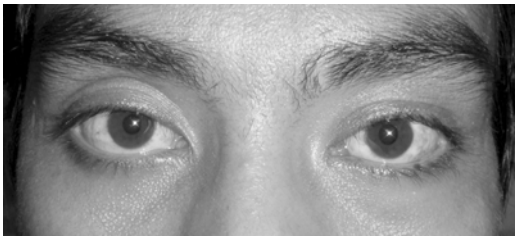

Figure 4 The left proptosis reduced after treatment.

function test results were normal. Collagen vascular and connective tissue disorder screening were negative. Culture and sensitivity was negative as well as the tuberculosis screening.

MRI of brain and orbit revealed generalized soft tissue swelling in the left extraconal and retro-orbital space suggesting of inflammatory reaction (Figure 3). The cavernous sinus and brain were normal. A chest radiograph was normal. Preliminary skin biopsy from the erythematous subcutaneous lesion was reported as 'benign panniculitis' (erythema nodusum). However, histopathology examination of a second skin biopsy revealed SPTCL. It was characterised by panniculitic-like pattern of infiltration by atypical lymphocytes surrounding the fat cells. Immunoperoxidase staining revealed positive cytotoxic T cell markers.

He was referred to the haematologist and was started on cyclophosphamide, hydroxy-daunomycin, oncovin and prednisone(CHOP) regime. At the same period, the intraocular pressure was regularly monitored and he was on eye lubricant to prevent exposure keratopathy. The patient responded well to the chemotherapy. The left eye proptosis and chemosis regressed(Figure 4). Visual acuity in both eyes was 6/6 and both fundi were normal. The left extraocular movement returned to full range. There was partial resolution of the subcutaneous lesions. Patient was followed up for about 12 months. He was healthy with no recurrence of proptosis.

DISCUSSION

SPTCL is a distinct type of peripheral T-cell lymphoma. It is a rare, malignant neoplastic proliferation of T lymphocytes that involves the subcutaneous fat lobules. This disease presents with a complex clinical, pathologic and immunohistochemical features. It is considered as an aggressive form of lymphoma. But, some patients manifest a long waxing and waning phase unaccompanied by constitutional symptoms. The incidence of SPTCL is very rare; being less than 1% of the non-Hodgkin lymphomas [1].

The disease appears clinically as non-specific multiple palpable subcutaneous nodules or deeply seated plaque most commonly in the extremities and trunk. Other less common sites of involvement are the face and back. Usually the subcutaneous nodules are painless and may regress spontaneously. Areas of lipoatrophy are seen in a regressed nodule. Rarely, ulceration develops over the nodules (6%)[1]. Lympahadenopathy and hepatosplenomegaly occurred in a minority of the patients. The patients present with the prodromal symptoms of unexplained high fever, chills, weight loss and myalgia[3]. More than 50% of patients have cytopaenia. Metastasis or visceral involvement is uncommon. Biopsies of the affected organ reveal extensive haemorrhage and histiocytes and phagocytising red blood cells.

Proptosis as a presentation of SPTCL is very rare. This patient initially presented with history of prolonged fever associated with generalized malaise and myalgia. The proptosis of the left eye appeared soon after that. This made the patient presented to the eye clinic first. Accompanying presentation were multiple erythematous subcutaneous nodules at various stages of healing. In the western counterparts, Leonard *et al*[4] had reported atypical case of SPTCL presented with diplopia and proptosis with bilateral abducens and superior and inferior oblique muscles palsies and active phagocytic activity. The patient succumbed to theillness despite chemotheraphy, allogenic transplantation and total body irradiation. Zhang *et al*[5] reported a case of SPTCL with unusual presentation of left eyelid swelling which was successfully treated CHOP regimen.

The most important investigation is histopathologic, immun-

ophenotypic and molecular analysis of the biopsied subcutaneous tissue. Histologically, small to large size lymphocytes are present in lobular pattern. The lymphocytes show atypical features such as hyperchromatic and angulated nuclei and indistinct cell borders. Rimming of fat cells by the neoplastic lymphocytes (CD_8) identified by immunohistochemistry is a diagnostic clue. However, presence of histiocytes, plasma cells and neutrophils may give false impression of benign panniculitis. Thus, repeated biopsy is needed if there is a clinical suspicion of lymphoma.

This patient later developed spiking temperature on admission. However, culture and sensitivity from blood, eye, throat, urine and urethra swab were negative. Full blood picture showed no presence of blast cell. Preliminary skin biopsy of the erythematous subcutaneous skin lesion in the patient reported as 'benign panniculitis'. However, second skin biopsy confirmed a diagnosis of SPTCL. As the aforementioned, the diagnosis was often difficult due to the histiologic similarities between the two entities and the indolent course of the disease. Perez-Persona *et al*[6] reported a case of SPTCL diagnosed after three skin biopsies within six months, by which time the lymphoma had become aggressive. A diagnosis of SPTCL may be difficult and often requiring repeated biopsy due to the indolent clinical presentation and histiologic similarities to benign panniculitis (erythema nodusom)[1,6]. As in this patient, only after the second skin biopsy, the diagnosis was revealed. The rare presentation of proptosis secondary to SPTCL had initiated early series of investigations and subsequently the final diagnosis. Standard treatment is by CHOP chemotherapy. Recent studies suggest long term remission is possible with high dose systemic steroid[1]. He was started on chemotherapy (CHOP regime) and responded well to the treatment. Reports in the literature describe remission in patients treated with CHOP chemotherapy regime[5,6]. So far there is no remission until the last follow-up about 12 months after the chemotherapy.

Aggressive cases not responding to initial immnunosuppressive therapy are treated with anthracycline-based and anthracedione-based chemotherapy combine with cyclopho-sphamide, hydroxydaunomycin, oncovin and prednisone. Radiation therapy is used for localised disease without haemophagocytic syndrome and palliative purposes. High-dose chemotherapy and stem cell transplantation resulting in high complete remission rate has been reported[6]. Poor prognosis and fatality are due to accompanying haemophagocytic syndrome which is a proliferation of hystiocytes and phagocytosis of blood element, hepatosplenomegaly and coagulopathy[1,3]. Under WHO-EOTC (World Health Organization-European Organization for Research and Treatment of Cancer) classification of cutaneous lymphoma, SPTCL is restricted to cases that express TCRαβ phenotype which carries a favourable outcome. In this type, haemophagocytic syndrome is uncommon (17%). There is a long clinical remission. 5 years survival rate is 82%. Long-term control can be achieved with systemic corticosteroid.

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急性突眼发作继发于皮下脂膜炎样 T 细胞淋巴瘤 1 例

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摘要

目的:报告罕见突眼急性发作病例继发于皮下脂膜炎样 T 细胞淋巴瘤(SPTCL)1 例。

方法:病例报告。

结果:患者,男,27 岁,马来人,左眼眼球突出急性发病 2wk。伴随长期高烧病史。在此期间他的身体和双大腿还多发肿胀红斑。双眼视力为 6/6。左眼眼球突出和球结膜水肿。各个方向眼外运动受限。角膜和眼前段正常。眼底检查显示正常视盘和视网膜。右眼检查为正常。体温 40.0℃,全身多发皮下红斑病灶主要分布在左腋下、右季肋部、双大腿和耻骨上区。腹股沟淋巴结也是可触及的。脑磁共振成像显示左冠外和眶后广泛软组织肿胀,暗示着炎症反应。海绵窦和大脑正常。红斑病灶处皮肤切片揭示为 SPTCL。其被提交给血液病医师,并开始 CHOP 方案化疗。患者化疗反应良好,眼球突出复位。

结论:眼球突出继发于 SPTCL 是非常罕见的。这是一个外周 T 细胞淋巴瘤的变异,特点是多发皮下结节表现为眼球突出和发烧。

关键词:皮下脂膜炎样 T 细胞淋巴瘤;良性脂膜炎;突眼