Case Report

Tenosynovial giant cell tumors an unusual presentation

Tahir Iqbal Mirza (BSc,MBBS,FCPS), Fasiha Tahir, Muhammad Umar Zahoo (FCPS), Mubshra Sameena (FCPS), Muhammad Ali Khan (FCPS), Mahmood Ahmed (FCPS),

1Surgical Department FC Hospital Quetta.  
2Gynaecologist FCH Quetta. 
3Asst Prof QIMS and Consultant Anesthesiologist and HOD FC Hospital.  
4Surgical Department CMH Mardan  
5Rehabilitation Medicine CMH Malir

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Tenosynovial giant cell tumors (TGCT) are barely malignant soft tissue tumors of the tendons, synovial bursas and joints. They are classified into two, common localized type and rare diffuse type. TGCT have been termed as nodular tenosynovitis, villonodular synovitis (pigmented) tenosynovitis and bursitis. Localized types commonly exist in finger joints while subtypes of diffuse-type are found in intra-articular or extra-articular places. The rare diffuse form is considered the soft tissue counterpart of diffuse pigmented villonodular synovitis and typically affects the lower extremities. Excision is the choice of treatment and recurrence is common feature. This is a case of 2nd toe right foot with unusual presentation of itching and treated dermatitis. It was managed with excision and follow-up in surgical OPD.

Key words: TGCT, Itching, Unusual presentation, barely malignant, excision, recurrence, disease modifying agents.

INTRODUCTION

The primitive description was endorsed by Chassaignac in 1852, which described these benign soft-tissue masses and overstated their biologic potential in referring them as cancers of the tendon sheath. While first elaborated description was published on 1941, by Jaffe et al. (Vasconez et al., 2007; Choudhury et al., 1999). Usual presentation is mass, local discomfort and pain, but no itching. The lesion may appear anywhere in the synovium, but in 80% to 90% of cases, it occurs in the hand joints, and infrequently in the knee and foot joints (John et al., 2000; Taylor et al., 2003; Buchner et al., 2005).

Higher incidences are not specific to any age group or gender. Studies described by Somerhausen et al. (2000), showed a little edge of women in prevalence (28:22). Pathogenesis of TGCT is yet not clear; they are benign but somehow included in semi malignant tumors because of their high recurrence rate up to 50% (Rao and Vigorita, 2000; Kuhnen et al., 2005). Sequence of events develops with fibroblastic activity, followed by histocytic proliferation, later on phagocytosis of RBGs, turns them in to haemosiderin –packed macrophages and giant cells. Excision of tumor is the recognized treatment, but other means like use of disease modifying agent is also getting acknowledgment in recent clinical trials (Cupp et al., 2007).

Case report

A 34-year-old mother of two, Pakistani settler in Afghanistan reported at Frontier Corps Hospital Quetta with complaint of severe itching and ill-defined mass at planter aspect of second toe right foot (Figure1). She was diagnosed as a case of dermatitis. The skin of right fore foot was without any scaring or pigmentation. There was a mass about 3 x 1.8cm at the area of proximal phalanx of the second toe, which was firm with some mobility along the long axis, mobile joints, preserved
Figure 1. Itchy mass about 3x1.8 cm at planter aspect right 2nd toe (TGCT).

neurovascular status and no lymphadenopathy. X-rays showed no bony lesion. FNAC results were Grade C-1 lesion. Excision biopsy was planned. With regional block, a vertical incision about 4 cm was made, at planter aspect of second toe. An adherent mass with tan white colour with size about 3x2 cm involving flexor sheath was noted. Instead of so deep invasion of the sheath flexor digitorum longus and brevis were spared. The tumor was in total cleared; with sparing tendon of flexor digitorum brevis. The recovery was uneventful.

Histopathology report from Agha Khan University Hospital Karachi read as fibrocollagenous tissue with aggregates of polygonal cells having variable collagenous depositions, moderate cytoplasmic changes and oval to round nuclei (Figure 2). An admixed lymphocytic infiltration also noted. Lesion also sounded positive for CD68 immuno-histological stain, while desmin, cytokeratin CAM- 5.2 and CD-138 were negative. On follow up she was observed symptom free (no more itching) with no residual mass on X-rays or MRI after one and half years.

DISCUSSION

TGCT is a rare pathological entity affecting synovium in young adults. Initially believed to be an inflammatory reactive process but researches in recent era have shown that this disease may actually be a benign neoplastic process with genetic alteration. Indeed a specific t(1;2) translocation, involving the collagen 6A3 gene(on 2q35) and macrophage colony-stimulating factor (M-CSF) gene(1p13) is present in a fraction of tumor cells in TGCT. Translocation involvingCSF1 at 1p13 is responsible for this clonal neoplastic proliferation through a landscape effect (Cupp et al., 2005; West et al., 2006). The clinical presentation of this tumor remained a mystery for diagnosis; it can be without any symptom or extreme numbness (Findling et al., 2011).

TGCT may presents with discomfort, heaviness, pain, swelling, limitation of movements in the affected joint. But itching is not a usual presentation, like in our case. She presented with sever itching, no pain or limitation of movements. Patient remained on continuous use of anti-allergic and steroid drugs both locally and systemic for 8 to 10 years. Such symptom of TGCT has not been quoted in medical literature but few post surgical excision cases were reported burning sensations in feet (Somerhausen and Fletcher, 2000; Kuhnen et al., 2005; Cupp et al., 2005; West et al., 2006; Hensley, 2010).

In our case no such symptom has been narrated by the lady, even after excision of the tumors mass. An important characteristic of the tumor is its slow growth, which leads to its usual diagnosis only by coincidence.
Differential diagnosis like lipoma, ganglia or fibromas should also be considered. Prior to an operation, it is difficult to distinguish between benign and malignant tumor. There is still some ambiguity in the etiology and no definitive research based evidences are available, as our patient told a history of small cut scar in childhood. Surgery is the main stay but Imatinib showed promising activity in recurrent episodes (Hensley, 2010).

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REFERENCES