

## ATYPICAL FIBROXANTHOMA IN TUBEROUS SCLEROSIS

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Atypical fibroxanthoma is not an uncommon condition occurring in elderly on the sun exposed areas. We report a case of atypical fibroxanthoma in a patient with tuberous sclerosis. The lesion was excised surgically. There was no evidence of any metastasis.

**Key Word : Atypical fibroxanthoma**

### Introduction

Atypical fibroxanthoma of the skin, a common tumour, was first described by Helwig in 1963.<sup>1</sup> It is now regarded as a low grade malignant tumour.<sup>2</sup> We report a case atypical fibroxanthoma in a patient with tuberous sclerosis.

### Case Report

A 56-year-old man was admitted in our ward with multiple facial fibrous papules and history of epilepsy. He was an old case of tuberous sclerosis who was admitted several times over the past 30 years.

During the course of his stay in the ward he developed a firm asymptomatic nodular swelling of 2.5 cm in size over the front of the neck. The skin over the swelling was shiny and stretched. There was no ulceration or granulation over the swelling.

An excision biopsy was done and the biopsy specimen showed a dermal infiltrate throughout the dermis impinging on the subcutis. Some cells were spindle-shaped with hyperchromatic nuclei and other cells were large and resembled histiocytes. Large bizarre-looking multinucleated giant cells were seen in the fibrous stroma (Fig.1). A diagnosis of atypical fibroxanthoma was made. There was no regional lymphadenopathy.

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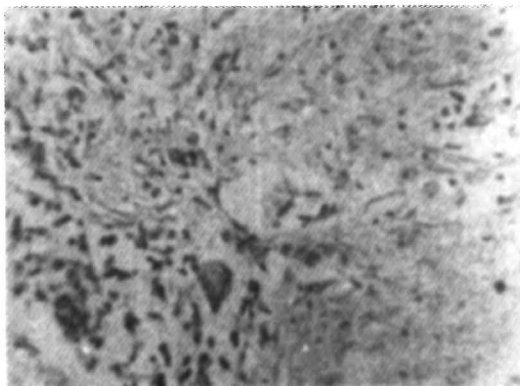


Fig. 1. Photomicrograph showing a large bizarre giant cell (H&E x 200).

Immunohistochemical studies for alpha-1-antitrypsin was not done for want of facility. Later he developed scrotal gangrene and lost contact with us.

### Discussion

Atypical fibroxanthoma was first thought of as a reactive tumour.<sup>1</sup> But there were reports of metastasis to the regional lymph node<sup>3</sup> and elsewhere.<sup>4</sup> Atypical fibroxanthoma is a raised nodular lesion occurring over the sun-exposed areas mostly head and neck in elderly and may be ulcerated.<sup>5</sup> Sometimes this can arise from x-ray irradiated areas.<sup>6</sup> In our case the nodule occurred over the neck of known case of tuberous sclerosis, a dominantly inherited disorder known for its association with various fibrous tumours of the skin like angiofibromas, shagreen plaque and multisystem tumours. Even though he underwent occasional roentgenography, there was no history of any irradiation. But our

patient is a farmer who is prone for prolonged sun exposure. So, the combined factors of genetic proneness and actinic damage might have been the cause in this patient. On review of literature there was no reported association between tuberous sclerosis and atypical fibroxanthoma.

## References

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