Answer to Dermacase continued from page 519

3. Atypical fibroxanthoma

Atypical fibroxanthoma (AFX) is a cutaneous tumour of fibrohistiocytic origin, which typically presents as a solitary, firm, dome-shaped nodule. It is usually well demarcated. The lesion grows rapidly but tends to remain less than 2 cm in size.1 Ulceration and minor bleeding can occur. The colour usually ranges from pink to erythematous, although a pigmented variant has been described of a brown-bluish colour resulting from hemosiderin deposition.² This pigmented form can easily be mistaken for melanoma.

Atypical fibroxanthoma is primarily seen in the headand-neck region of the elderly (mean age in the seventh decade). The nose, cheeks, and ears are the most common sites affected.3 Occurrence on the scalp might also be more common than previously recognized. It is possible AFX might arise on the trunk and extremities, in which case the lesions tend to be larger, less well demarcated, and grow more slowly compared with those that appear on the head and neck.4 This distribution, however, is infrequent and predominantly occurs in a younger population of patients (mean age in the fourth decade).3 Atypical fibroxanthoma is most prevalent in the white population, and men tend to be affected more than women 4,5

The main predisposing factor is prolonged exposure to sunlight, supported by the fact that AFX frequently appears on a background of sun-damaged skin. In addition, most patients have multiple actinic keratoses and a past history of basal cell and squamous cell carcinomas.5 Other possible predisposing factors include a history of radiation therapy and local skin trauma.⁶ The exact pathogenesis of AFX is not entirely understood.

Diagnosis

Establishing the diagnosis of AFX based on clinical grounds can be challenging; it is relatively uncommon and frequently resembles other entities. The clinical differential diagnosis includes basal cell carcinoma, squamous cell carcinoma, melanoma, keratoacanthoma, pyogenic granuloma, and epidermoid cysts.^{1,5,6} Therefore, a skin biopsy is essential in making the correct diagnosis. The histologic differential diagnosis includes squamous cell carcinoma, malignant melanoma, leiomyosarcoma, and malignant fibrous histiocytoma.⁵ Immunohistochemical staining performed by the pathologist helps differentiate these tumours.

Treatment

Atypical fibroxanthoma is considered a low-grade malignancy. It has a relatively benign course and carries a favourable prognosis with appropriate treatment. Metastases (local or distant) are rare and have been reported to occur in approximately 1% of cases.⁶



Atypical fibroxanthoma is treated surgically. Wide surgical excision with 1-cm margins used to be the treatment of choice; however, Mohs micrographic surgery is now the preferred method. Mohs surgery offers 3 advantages: lower recurrence rates, tissue conservation in important cosmetic areas such as the head and neck, and assurance of complete excision (tumour-free margins) while performing the surgery.^{5,6} Recurrence rates range from 0 to 7% with Mohs surgery, and up to 16% with wide excision.7 Owing to the possibility of the tumour extending into subcutaneous tissue and, rarely, metastasizing, cryosurgery and electrodesiccation and curettage are generally not recommended for treatment of AFX.1 Regardless of treatment, however, all patients should follow up within 6 months to ensure there is no local recurrence.

Mr Prajapati *is a third-year medical student at the* University of Calgary in Alberta. Dr Barankin is a dermatologist in Toronto, Ont.

Competing interests

None declared

References

- 1. Stadler FJ, Scott GA, Brown MD. Malignant fibrous tumors. Semin Cutan Med Surg 1998;17(2):141-52.
- 2. Diaz-Cascajo C, Borghi S, Bonczkowitz M. Pigmented atypical fibroxanthoma. Histopathology 1998;33(6):537-41
- 3. Anderson PJ, McPhaden AR, Ratcliffe RJ. Atypical fibroxanthoma of the scalp. Head Neck 2001;23(5):399-403.
- 4. Fretzin DF, Helwig EB. Atypical fibroxanthoma of the skin. A clinicopathological study of 140 cases. Cancer 1973;31(6):1541-52.
- 5. Seavolt M, McCall M. Atypical fibroxanthoma: review of the literature and summary of 13 patients treated with Mohs micrographic surgery. Dermatol Surg 2006;32(3):435-41.
- 6. Giuffrida TJ, Kligora CJ, Goldstein GD. Localized cutaneous metastases from an atypical fibroxanthoma. Dermatol Surg 2004;30(12 Pt 2):1561-4.
- 7. Huether MJ, Zitelli JA, Brodland DG. Mohs micrographic surgery for the treatment of spindle cell tumors of the skin. J Am Acad Dermatol 2001;44(4):656-9.