Dear Editor,

Acquired progressive lymphangioma (APL) is a rare benign lymphatic proliferation, typically arising as a solitary lesion on the trunk and limbs in both children and adults. It can appear flat, indurated or as a plaque and exhibits a range of appearances from red, brown, violaceous or yellow. Histological findings need to be distinguished from more aggressive entities and can be challenging without further history.

A 75-year-old man of Italian descent presented with an ill defined scaly, slightly indurated, erythematous lesion on the right upper chest (Fig. 1) associated with pruritus and occasional serous fluid discharge from minute breaks in the skin. It grew slowly over a 5-month period and subsequently remained unchanged. Recent swabs, inflammatory markers and autoimmune screen have been unremarkable. He had a normal full blood count, liver function studies, urea and electrolytes and had negative serology for Borrelia burgdorferi, anti-nuclear antibodies and antibodies to extractable nuclear antigens (ENAs).

Twenty-two years ago, he had undergone 2 orthopaedic procedures 3 months apart on the right shoulder under general anaesthesia. At the initial operation, a ganglion-like lesion had been removed from the right acromioclavicular joint area complicated by a discharging synovial fistula. This fistula was subsequently excised and the distal clavicle resected at the acromioclavicular joint. After the second surgery, the discharge stopped and the patient made a full recovery. Consistent with the surgical findings, histopathology of the dermis and underlying connective tissue from the second operation had revealed a fibrocollagenous stroma associated with a chronic inflammatory infiltrate and granulation tissue.

During biopsy on this occasion, a mucoid liquid was noted emanating from the biopsy sites. Histopathology revealed multiple dilated, thin-walled, endothelial-lined channels in the superficial dermis with some showing occasional intraluminal red blood cells and some markedly dilated lumina. A small amount of disorganised smooth muscle was seen in the walls of the larger spaces, and in one area, a very large lymphovascular structure was seen in the upper dermis. Some of the vessels lay parallel to the epidermis, and none showed endothelial atypia or mitotic activity. There was no multilayering or papilla formation. The endothelial cell lined spaces dissected through dermal collagen in areas with extension into deep dermis and some background fibrosis (inset).

Immunohistochemical staining showed the endothelial cells to be CD31 and CD34 positive. The lymphatic endothelial marker D2-40 (podoplanin) was strongly positive (not shown). These findings support a lymphatic origin for the lesion, in keeping with a lymphangioma. MRI of the right shoulder and anterior chest wall revealed a joint effusion extending into the subacromial bursa with associated synovitis, consistent with advanced glenohumeral arthropathy. There were no evidence of a soft tissue mass, communication or synovial leak to the skin. Due to the large size of the lesion and lack of symptoms, no further surgery was performed. The patient has been advised to avoid trauma to this area to reduce the chance of lymphorrhoea and secondary infection. It had remained unchanged at 12 months after first presentation.

Previous authors have found APL to develop after surgery, trauma or radiation therapy and propose it to be an inflammatory rather than neoplastic process. The
presentation is often subtle and generally asymptomatic, however, a discharge can occur secondary to scratching, trauma or infection. Furthermore, pruritus, tenderness and swelling have also been reported. Symptom duration may be variable and there is often a delayed onset from any insult up to 20 years. In this patient, the clinical differential diagnosis was diverse; including lymphangioma, lymphangiosarcoma, secondary deposits from a mucoid carcinoma, granuloma annulare, plaque-type mucinosis, lepromatous leprosy, sarcoidosis, deep fungal infection, erythema chronicum migrans and mycosis fungoides. Histology is essential to make the correct diagnosis.

Histologically, the superficial dermis is partly replaced by anastomosing, angulated and widely dilated vascular spaces, sometimes arranged horizontally, parallelizing the epidermis. These vascular spaces often dissect the dermal collagen and tend to become narrower, angular and cleft-like at deeper levels. There is an absence of atypia or mitotic figures and the spaces contain proteinaceous material, a few red blood cells or are empty. A smooth muscle component has also been demonstrated focally around the vascular spaces.

Immunohistochemical studies in APL have been inconsistent. Endothelial cells have been reported to stain positively and negatively for factor VIII-related antigen and Ulex europaeus-1 lectin (UEA-1) and positively for CD34 and CD31. The use of the lymphatic-specific stain D2-40 (podoplanin) is highly valuable in identifying the lymphatic nature of the proliferation, since it does not stain vascular endothelium. It has only recently become widely available, so could not be used to assess lesions in many previous studies.

When Wilson Jones first described APL, he differentiated it from angiosarcoma by these features: (1) young age of onset; (2) location away from the face and scalp; (3) a localised, flat pattern of growth; (4) slow growth and favourable prognosis; (5) dissection of collagen by the endothelial-lined channels without cellular atypia. Histologically, the differential diagnosis includes malignant conditions such as well-differentiated angiosarcoma and patch stage Kaposi’s sarcoma: clinicopathologic analysis of a series. Am J Surg Pathol 2000;24:1047-57.

The aetiology of APL is uncertain, but it appears to be a benign lymphatic proliferation, possibly a response to prior dermal injury. Awareness of this condition is important to avoid misdiagnosis for conditions such as AS or KS. The use of D2-40 is helpful to recognise the true lymphatic origin of the lesion. Where it is practical, smaller lesions are best managed by surgical excision and have an excellent prognosis.

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REFERENCES


Philip L Tong, MBBS (Hons), Trevor W Bee, MBChB, MRCPath, FRCPA, Daniel Fick, MBBS (Hons), FRACS, Sujith P Kumarasinghe, FCCP, FAMS, FACS

1Sir Charles Gairdner Hospital, Perth, Australia
2Cutaneous Pathology, Nedlands, Perth, Australia
3Department of Orthopaedic Surgery, Sir Charles Gairdner Hospital, Perth, Australia
4Department of Dermatology, Royal Perth Hospital, Perth, Australia

Address for Correspondence: Dr Philip Tong, Sir Charles Gairdner Hospital, Hospital Avenue, Nedlands 6009, Western Australia.
Email: philipleitong@gmail.com