Adolescent breast lymphoma - apparently aggressive presentation with favourable outcome

Sir,

Primary breast lymphoma (PBL) is a rare entity making up less than 0.5% of breast malignancies. The occurrence of PBL in adolescence is rare. Anaplastic large cell lymphoma (ALCL) is an unusual variant with apparently aggressive features. We report such a case with a very favourable long-term outcome.

A 15-year-old girl presented in 1981 with a 5-cm ulcerating lump in her left breast of five months duration (Stage 5 adolescent development). There was no axillary lymphadenopathy (Figure 1a). Wide local excision with split skin graft coverage was done (Figure 1b). Grossly, a 5-cm mass of ulcerating pale tissue was seen. Microscopically, sheets of malignant “epithelial” cells with anaplastic arrangement, conspicuous pleomorphism and mitoses were seen (Figure 2a) which was interpreted as a medullary or encephaloid carcinoma. She was treated with oral cyclophosphamide for 31 months and prednisolone for seven months. Neither adjuvant radiotherapy nor tamoxifen was offered. She remains well with two healthy children at 22 years of follow-up.

Review of the histology was done as the patient was referred to our hospital. The blocks were preserved and further sections were cut. Immunostaining was performed using standard procedures with a streptavidin-biotin detection system. Heat-mediated antigen retrieval was used where appropriate. Immunocytochemistry showed that the cells failed to stain with antibodies to cytokeratin (CAM 5.2, AE1/AE3) (Figure 2b) and S100 protein. There was focal staining for epithelial mem-

Figure 1a: Clinical presentation demonstrating that the tumour arose in the breast

Figure 1b: After wide local excision and skin grafting

Figure 2: (a) The H&E stained section shows a sheet of large cells with abundant cytoplasm and moderate nuclear pleomorphism. (b): The cells do not stain for cytokeratin (c): The cells express CD30 (d): The cells express ALK-1
brane antigen. The cells stained strongly for CD30 (Figure 2c) with positive staining for CD45 in some cells. The cells were negative for T cell, B cell and NK cell markers. The appearances were consistent with the diagnosis of anaplastic large cell lymphoma (ALCL) of the breast of common type with a null phenotype. Staining with ALK-1 gave a characteristic nuclear and cytoplasmic pattern seen in cases with the t(2;5) translocation associated with this tumour (Figure 2d). Immunostaining for oestrogen receptor (ER), progesterone receptor (PgR) and Her 2 were negative.

Breast tumours during adolescence are most frequently benign. The rarity of PBL is probably related to the paucity of natural lymphoid tissue in the breast. Diagnostic criteria for PBL set by Wiseman and Liao\(^1\) included 1) technically adequate material for review; 2) the breast as the primary tumour site and 3) no prior documentation of a similar histological type of lymphoma elsewhere. The majority of ALCL cells stain positive for the CD30 and anaplastic large cell lymphoma kinase (ALK) protein, which may also be a useful prognostic factor.\(^2\) The overall 5-year survival rate in ALK-positive tumours is almost 80% in contrast to only 40% in negative tumours. However, these tumours often remain sensitive to chemotherapy. Our patient had an ALK-positive tumour and this probably predicted her long survival. Our patient is of the common variant type according to the Stein\(^3\) subclassification of ALCL.

As PBL is rare, clinical management protocols are often based on the treatment of lymphomas in general. The most common chemotherapy combination for non-Hodgkin lymphoma and non-specified malignant lymphoma is based on the CHOP regime consisting of cyclophosphamide, doxorubicin, vincristine and prednisolone.\(^3\) Although ALCL is associated with good prognosis, long-term follow-up is required. Recurrence may be loco-regional, in the contralateral breast\(^4\) or systemic and occurs in about 30% of cases.

Women presenting with rare breast lymphomas should be accurately diagnosed and the subtype characterised to facilitate optimum treatment and prognosis. ALK-positive ALCL are associated with good prognosis.

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References