with an aggressive course and are characterized by an angiocentric, angiodestructive, lymphoid infiltrate. Immunohistochemically these cells are $CD2^+$, $CD3^-$, $CD7^{+/-}$, and $CD56^+$.

In general, $CD56^+$ tumors have a poor prognosis, with a mean survival of 13 months.[4] It has been reported that lymphomas that overexpress the p53 gene have a worse prognosis.[8]

These tumors must be differentiated from blastic NK-cell lymphoma. This type of tumor was considered to arise from immature NK cells, but it is now known that the precursor cells are plasmacytoid dendritic cells.[9]

Due to their rarity, it is important to recognize the existence of highly aggressive lymphomas and to classify them correctly. Both pathologists and clinicians must keep these tumors in mind, make the correct diagnosis, and start specific treatment rapidly. Radiation therapy is the treatment of choice in localized disease and is usually combined with chemotherapy.[3] If patients are initially managed using chemotherapy, palliative or coadjuvant radiation therapy is recommended, as its benefits persist even after chemotherapy.[10]

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Paraneoplastic Ichthyosis

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To the Editor:

We report the case of a 63-year-old woman with a history of type 2 diabetes and hyperuricemia who presented with asthenia, weight loss (totalling 14 kg), pruritus, xerosis, and diffuse scaling. The symptoms had begun 4 months earlier. The patient also had a growth under her right arm that had been present for a month.

On examination, the patient was found to have marked xerosis and widespread scaling, with small, loosely adherent, confluent scales that were whitish in color. The most severely affected areas were the extensor surfaces of the arms and legs; here the flakes were browner and there were also reticulated lesions reminiscent of eczema craquelé (Figure 1). Scaling was also evident on the flexor surfaces and on the face and scalp. Palpation revealed enlarged axillary lymph nodes under both arms (the node in the right axilla measured 5 cm) (Figure 2). Computed tomography showed masses in the mediastinum and in both

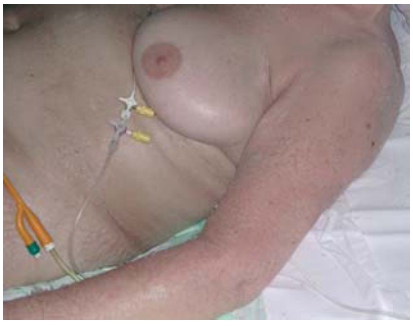

Figure 1. Small, widespread, partially adherent, whitish scales.

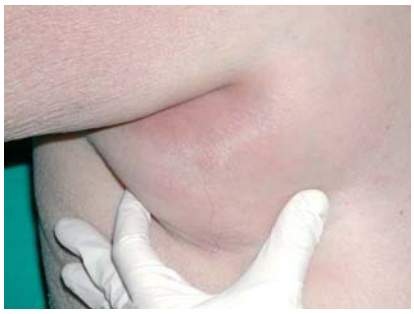

Figure 2. Subcutaneous mass in right axilla.

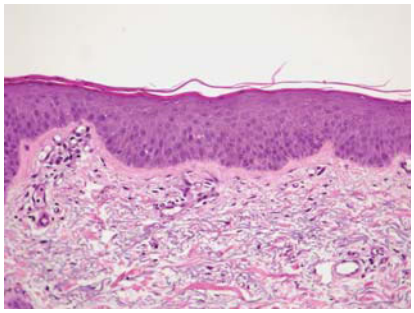

Figure 3. Parakeratosis and absence of the stratum granulosum (hematoxylin–eosin, original magnification ×200).

axillary regions. Histology of the right axillary lymph node confirmed the diagnosis of stage IIB nodular sclerosing Hodgkin disease. The findings of the skin biopsy were consistent with paraneoplastic ichthyosis, with loss of rete ridges, parakeratosis, and practically no detectable stratum granulosum (Figure 3).

The disease continued to progress despite initiation of polychemotherapy. The cutaneous symptoms also worsened as the lymphoma progressed, despite treatment with topical emollients and corticosteroids. The patient died 18 months after diagnosis.

Acquired ichthyosis is much less common than its congenital counterpart. The acquired form of the disease has been associated with many systemic diseases, including cancer, leprosy, sarcoidosis, thyroid disorders, hyperparathyroidism, diabetes, chronic kidney failure, effects of bone marrow transplant, and human immunodeficiency virus infection.[1-3] The condition has also been linked to nutritional disorders and autoimmune diseases, such as systemic lupus erythematosus and dermatomyositis, as well as to the administration of drugs such as niacin, triparanol, butyrophenones, cimetidine, and chlofazimine.[1-3] The most common underlying disease (and also the most serious in terms of prognosis) is cancer, the diagnosis of which tends to coincide with the manifestation of the clinical signs of ichthyosis. Acquired ichthyosis is most often associated with Hodgkin disease, but it has also been linked to non-Hodgkin lymphoma, Kaposi sarcoma, leiomyosarcoma, and cancers of the breast, lung, ovary, and cervix.[1-3]

Acquired ichthyosis is very similar to ichthyosis vulgaris, both clinically and histologically, and some authors have actually classified the acquired disease as acquired ichthyosis vulgaris.[1,4] Although the extensor surfaces of the arms and legs were the most severely affected areas in our patient, there were also lesions on the face and the flexor surfaces, ruling out hereditary ichthyosis vulgaris. While the histology of paraneoplastic ichthyosis is not specific, remarkable findings include foci of parakeratosis and a thin or absent stratum granulosum.

Although paraneoplastic ichthyosis normally runs a parallel course with the underlying cancer, it may also appear earlier or later. The skin condition improves with treatment of the cancer and the return of lesions may indicate tumor recurrence.[4,5] When the tumor is incurable, ichthyosis can be controlled using emollients, topical or systemic corticosteroids, or retinoids.[1,4]

Not a lot is known about the pathogenesis of paraneoplastic ichthyosis, but it has been suggested that reduced lipogenesis in both the dermis and epidermis might play an important role.[6] One proposed model for the pathogenesis of paraneoplastic dermatosis included 3 stages[7]:

1. Development of the tumor
2. Production of an inducer by tumor cells
3. Development of epithelial cell susceptibility to the inducer Accordingly, it has also been suggested that transforming growth factor-α produced by the tumor might be responsible for the proliferation of susceptible epithelial cells.[4,5]

In patients with adult-onset ichthyosis and no history of the hereditary form of the disease, we recommend taking an exhaustive history and conducting an analytic study and full clinical examination to rule out underlying internal disease, and cancer, in particular.

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