QUIZ 1

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Clinical and histopathological findings:

A white 50-year-old patient reported to the hospital because of a tumor on the right foot of one year duration. The tumor was firm, painless, moving underlying tissues and covered by normal skin (Figure 1).

This tumor had been rapidly growing for the past six months. Complete physical examination showed a large non inflammatory inguinal lymphadenopathy (Figure 2). A biopsy of the tumor showed the histopathologic findings shown in Figures 3 and 4.

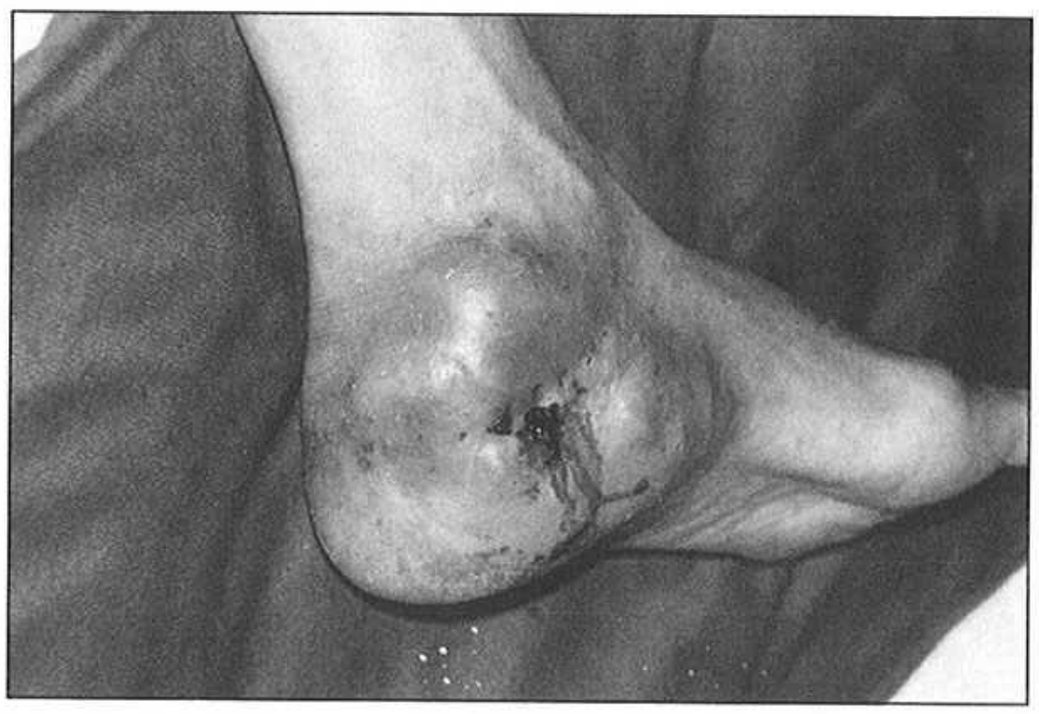

Fig. 1: Nodular tumor of the foot.

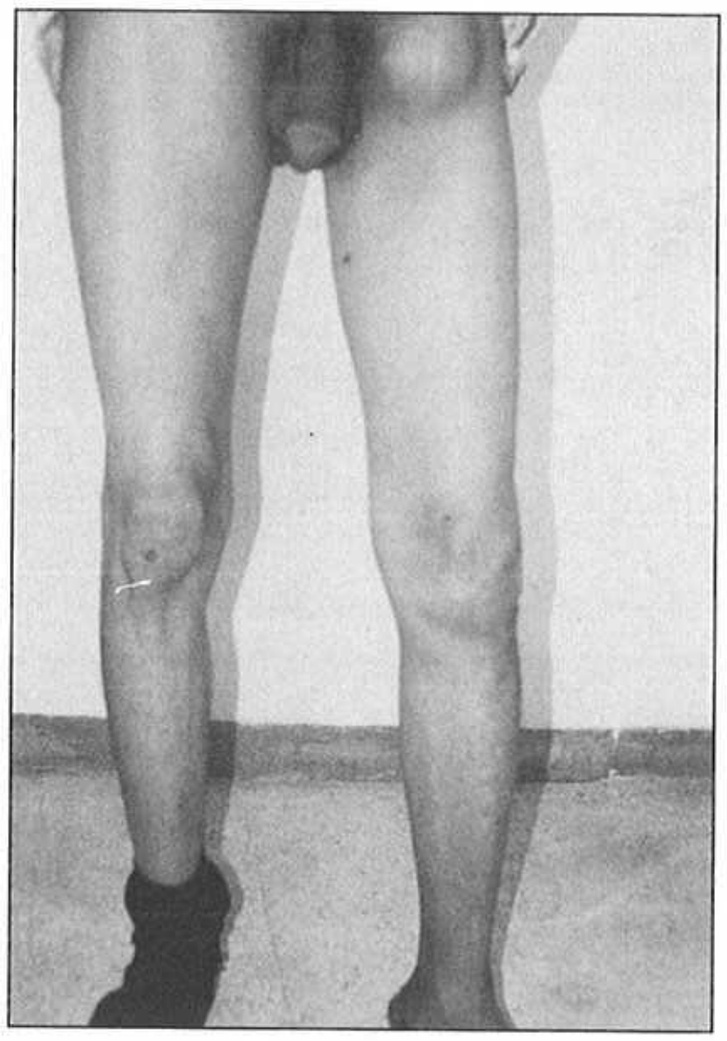

Fig. 2: Inguinal lymphadenopathy.

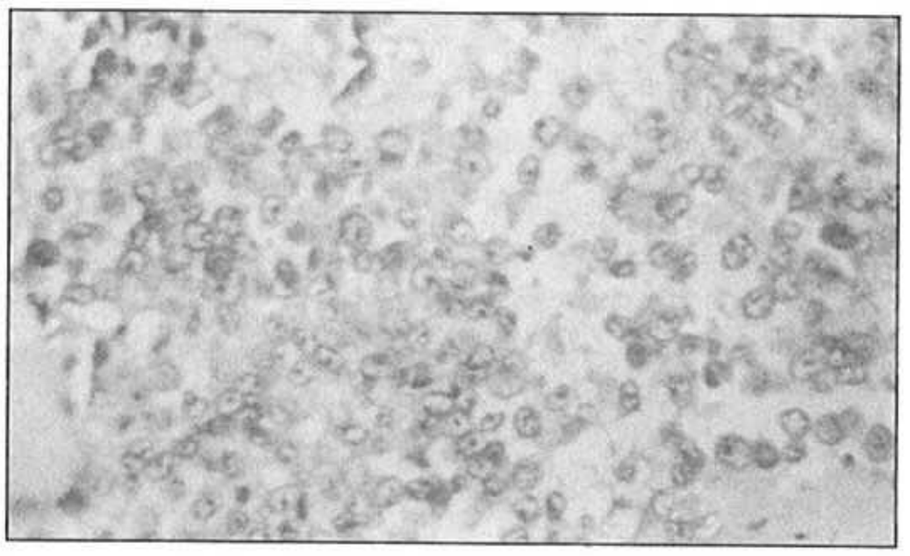

Fig. 3: Histopathologic findings, H.E X 250.

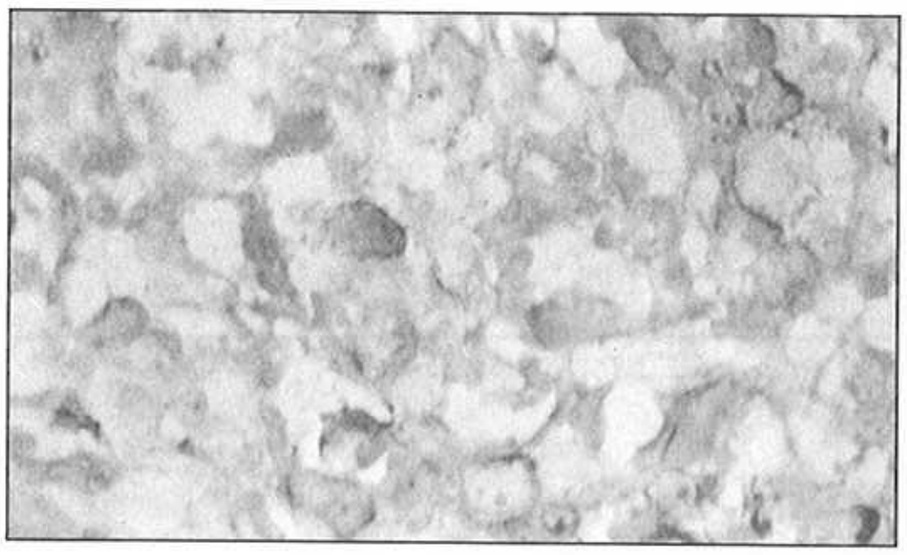

Fig. 4: Histopathologic findings, Grumelus X 400.

What is your diagnosis ?

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DIAGNOSIS :

Primary Neuroendocrine Carcinoma of the Skin

DISCUSSION

Primary Neuroendocrine Carcinoma of the Skin (PNCS) is a rare tumor described for the first time by Toker in 1972 as a trabecular carcinoma of the skin[1].

This tumor tends to occur in the elderly[2,3]. The sites of predilection are: the head neck (46%), legs (27%), arms (17%), and trunk (10%). Vulvar and anal sites were also reported[2,3,4,5].

The clinic features include: a painless nodular tumor, which might be covered by a violaceous, pigmented, ulcerated or sometimes normal skin.

Local recurrence is seen in 1/3 of the cases, a lymph node metastases in 1/2, and a visceral metastasis in 28 % of the cases[1,2,3,4].

The histologic study (light microscopic) reveals that the tumor proliferation is located within the dermis and extends to involve subcutaneous tissue. The growth consists of solid sheets of round cells or cells arranged in anastomosing cords and trabecular pattern. The tumor cells show a narrow amphophytic cytoplasm surrounding a vesicular nucleus that contains multiple nucleoli. Mitotic figures are abundant[6].

The electron microscopic study shows a dense core neurosecretory granules, measuring up to 150 nm diameter situated within blunt-ended cytoplasmic processes[7].

Recent advances in immunohistochemistry permitted to define the tumor characteristics with high precision. Many markers are used but some are more specific than others. The most important are the markers of the neuron-specific enolase (+) chromogranin A (+), protein S100 (-), leukocyte common antigen (-), and low molecular weight cytokeratine (+)[2,3,5,7].

This study is necessary to differentiate PNCS from other malignant tumors such as lymphoma, melanoma, and metastatic oat cell carcinoma.

It can be associated with other malignant processes such as: chronic lymphoid leukemia, skin tumors (basal cell carcinoma, epidermoid carcinoma, actinic keratosis), and Hodgkin disease[3,8].

To prevent the local recurrence of the tumor, it has to be excised together with the local lymphadenopathy[3]. The radiotherapy[9] and the chemotherapy[4,8,10] can be used as an adjunct to surgery or as a palliative treatment.

In conclusion, It is important to emphasize that the earlier the diagnosis, the better is the prognosis for this malignant tumor in which the affiliation with Merkel cell has been proposed but not confirmed[7,11].

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