Masson's papillary endothelial hyperplasia mimicking a poorly differentiated thyroid carcinoma: a case report

Annette Winkler, Bruno Mueller and Peter Diem

Division of Endocrinology and Diabetes, University of Bern, Inselspital, CH-3010 Bern, Switzerland

(Correspondence should be addressed to P Diem; Email: peter.diem@insel.ch)

Intravascular papillary endothelial hyperplasia (PEH, Masson's tumour) is a benign lesion of vascular origin that is caused by an excessive and atypical proliferation of endothelial cells in normal blood vessels or vascular malformations. PEH may mimic angiosarcoma or other tumours (1). On only very rare occasions does PEH occur extravascularly as a result of the organization of a haematomata (2), although a history of trauma is seldom elicited. There are only two reported cases in the literature of PEH of the thyroid, one occurred subsequent to multiple fine-needle aspiration biopsies, the second occurred in an organizing haematomata within a hyperplastic follicular nodule (3, 4). We present here what we believe to be the first report of a patient with PEH of the thyroid that was mistaken for a poorly differentiated thyroid carcinoma on the original histology.

A 45-year-old woman presented in 1988 with a non-toxic multinodular goitre. Her medical history revealed no significant, pre-existing disease and her sister also suffered from goitre. Because of neck discomfort, the patient underwent subtotal thyroidectomy. Histological examination revealed foci with hyperplasia of the epithelial cells, colloid-rich adenomatoid nodules with haemorrhagic and cystic degeneration. Furthermore, an area with round to spindle-shaped cells with a high mitotic activity and infiltrative expansion exceeding the thyroid capsule was detected. Results from an immunohistochemical analysis of this area were negative for synaptophysin and calcitonin. The histopathological features were interpreted by an approved pathologist as a poorly differentiated thyroid carcinoma. Complete thyroidectomy was therefore carried out; total ablation with iodine-131 was not performed. Suppressive levothyroxine therapy was initiated. One year postoperatively, a thyroid radionuclide scan in combination with an ultrasound revealed an isolated 50 mm recurrent mass on the left side; lymph node metastases were not detected. Results of an iodine-131 total body scan were negative. Serum thyroglobulin concentrations were elevated (122 ng/ml; reference range <25) but calcitonin values were within the normal range. Apart from a follow-up including serum thyroglobulin and calcitonin measurements no further steps were taken.

In 1996 serum thyroglobulin concentrations increased rapidly (885 ng/ml). An ultrasound of the thyroid showed a 57 mm nodule with non-homogeneous echogenicity. Thyroid scintigraphy using 99Tc-pertechnetate and 99mTc-MIBI showed a positive tracer accumulation within this mass. Based on these findings, the patient was referred to our endocrine division, and the tumour tissue in the neck was removed. Histopathological evaluation revealed typical findings of a simple nodular goitre; there were no signs of malignancy. Because of the very unusual course of this tumour, the original specimens which had been conserved were histologically and immunohistochemically reanalysed. The spindle-shaped cells were found mainly to be endothelial cells with papillary proliferation within dilated venules. The additional immunohistochemical investigations showed positive staining to vimentin, to CD34 and to factor VIII-related antigen within the endothelial cells. Thus, histopathological examination revealed a pure form of PEH; the original diagnosis of poorly differentiated thyroid carcinoma was dismissed.

Intrathyroidal PEH is a very rare entity, but may have been under-reported in the literature (5). Because of its clinical course and histological findings, PEH may be mistaken for a malignant tumour, as is illustrated by our case. Misdiagnosis may lead to consequences such as inappropriate radical surgery. PEH can be cured by complete surgical resection. Our patient therefore underwent two unnecessary operations to remove the recurrent ‘malignant tumour’. It is also important to distinguish between these two lesions to avoid inappropriate medical treatment such as chemotherapy, total ablation with iodine-131 or levothyroxine-suppressive treatment. The latter has potentially serious side-effects, as has been described recently (6). The psychological aspect of our patient living with the diagnosis of malignant tumour must also be considered. Physicians caring for patients with thyroid disorders and pathologists should therefore be aware of PEH and suspect its presence.

References


Received 1 June 2001
Accepted 11 July 2001