Giant trichoblastoma of the scalp

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Trichoblastome géant du cuir chevelu

LA TUNISIE MEDICALE - 2011 ; Vol 89 (n°06) : 569 - 572

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RÉSUMÉ

Prérequis : Le trichoblastome est une tumeur annexielle bénigne rare avec un aspect histologique évocateur. Elle survient sur les régions pileuses et se présente habituellement sous forme d'une lésion solitaire de moins de 2 cm de diamètre. Le trichoblastome dans sa forme géante est une variante rarement décrite dans la littérature.

But : Rapporter un nouveau cas de trichoblastome géant déroutant vers la malignité.

Observation: une femme de 57 ans s'est présentée avec une lésion nodulaire unique et asymptomatique du cuir chevelu de 5 cm de diamètre évoluant depuis 28 ans. La lésion a été excisée à 2 reprises avec une récidive et reprise évolutive. L'examen a montré une lésion nodulaire érythémateuse, ferme et papillomateuse, non infiltrée en dôme. Le bilan d'extension était négatif. La biopsie cutanée a conclu à un trichoblastome mais n'a pas pu tranché quand à la bénignité de la lésion. Après excision greffe, l'histologie de la pièce a confirmé la bénignité et la totalité de l'exérèse. Il n'y a pas eu de récidive après 5 ans de suivi.

Conclusion: Cette observation, illustre une forme clinique rare de trichoblastome dit géant. Une néoplasie a été difficile à éliminer chez notre patiente, du fait de l'évolutivité de la tumeur. Cependant, l'évolution lente ainsi que la bénignité histologique ont permis de redresser le diagnostic. Cette tumeur est considérée par certains auteurs comme une entité à part entière.

Mots-clés

Trichoblastome géant ; trichoepitheliome

SUMMARY

Background: Trichoblastoma is a rare and benign adnexial tumor with characteristic histological features. It occurs on any hair follicle-bearing location, and usually presents as a solitary lesion most often less than 2 cm in size. Giant trichoblastoma has been rarely reported in the literature.

Aim: To report a new case of giant trichoblastoma, misleading for malignancy.

Case Report: A 57-year-old woman presented with a 5 cm-solitary asymptomatic nodular lesion of the scalp, of 28 years. It had been previously excised with recurrence and progressive regrowth. On examination, it was a dome-shaped, erythematous, firm, papillomatous, non infiltrated nodule. Full body work up revealed no metastases. Cutaneous biopsy concluded to trichoblastoma but failed to eliminate malignancy. After excision with secondary skin graft, histological examination confirmed the benignity with clear margins. There was no evidence of recurrence after a 5 year-follow-up period.

Conclusion: This case illustrates a rare clinical variant of trichoblastoma with an unusual important size. This can be misleading for malignancy, but the slowly progressive course of the tumour in our patient, together with histological benignity led to the correct diagnosis. This tumour is considered as a distinct entity by some authors.

Key-words

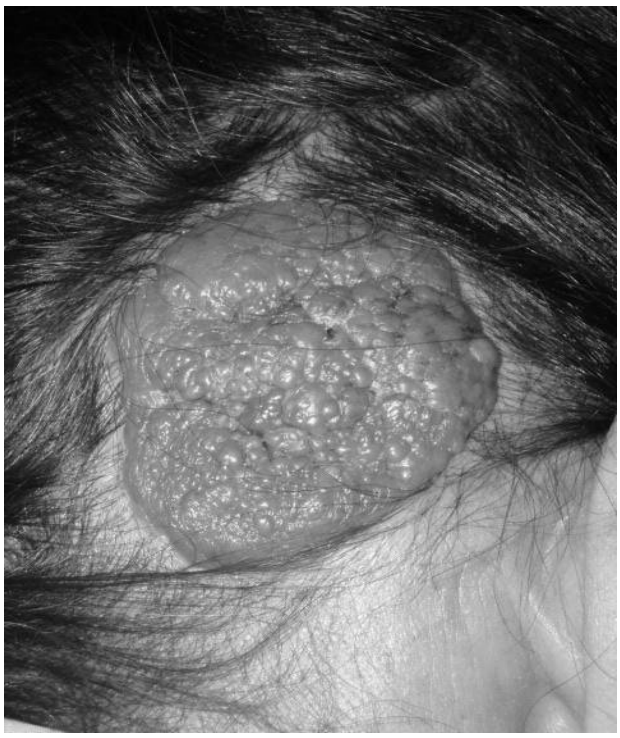
Giant trichoblastoma; trichoepithelioma

Trichoblastomas are benign adnexial tumors of follicular germinative cells [1]. They represent rare neoplasms composed of symmetric, well-circumscribed nests and lobules of small monomorphous basaloid cells with peripheral palisading, embedded in a fibrotic stroma. The germinative cells of the epithelial component often show follicular differentiation [2]. Different subtypes of trichoblastomas have been described including sebaceous and sweat gland differentiation, subcutaneous variants, pigmented trichoblastoma, and clear cell trichoblastoma [2 - 6]. They often appear as solitary little-sized lesions. But, trichoblastomas of larger and even gigantic size have been rarely reported [1, 7]. Through a case of giant trichoblastoma of the scalp we will emphasize the different clinical and pathological patterns of this adnexial tumor and its misleading clinical aspects when it becomes larger in size.

CASE REPORT

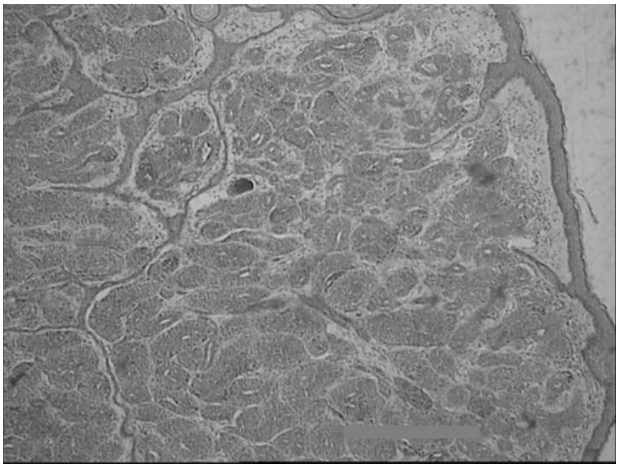
A 57-year-old woman presented with a solitary asymptomatic lesion of the scalp evolving for 28 years. There was no pre-existent lesion. It had been previously excised in 1979 and in 1991, with recurrence and progressive regrowth. Examination revealed a dome-shaped, erythematous, firm, papillomatous, non infiltrated nodule measuring 5 cm in diameter, located on the right occipital region of the scalp (Figure 1).

Figure 1 : 5 cm-dome-shaped, erythematous, firm, papillomatous, non infiltrated tumor of the right occipital region of the scalp.



There was no palpable lymph node. Histological examination of a cutaneous biopsy had concluded to trichoblastoma but the benignity could not be confirmed. Cranial CT scan showed no infiltration of the occipital bone and full body work-up revealed no signs of metastases. Deep tumoral excision, putting to naked the bone, was conducted with one cm margins. The second step consisted in total skin graft. Histological examination showed a well circumscribed but non-encapsulated nodular tumour of the entire dermis and the hypodermis (Figure 2A).

Figure 2A : A well circumscribed, non-encapsulated nodular tumour of the entire dermis



This tumour presented variably sized epithelial nests closely resembling basal cell carcinoma. Peripheral palisading was conspicuous and there was a stromal condensation around tumour lobules but cleft artefact was not a prominent feature (Figures 2B-2C).

Figure 2B : Large and small nests with peripheral palisading, surrounded by stromal condensation.

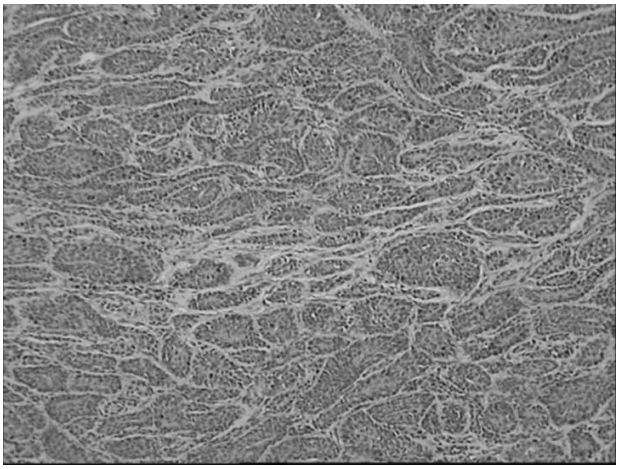
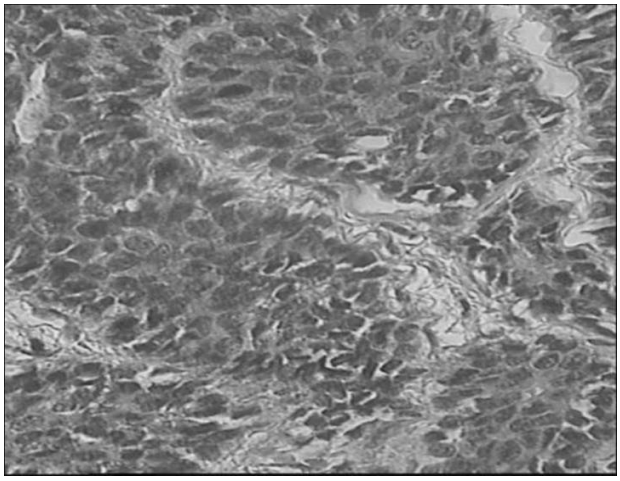
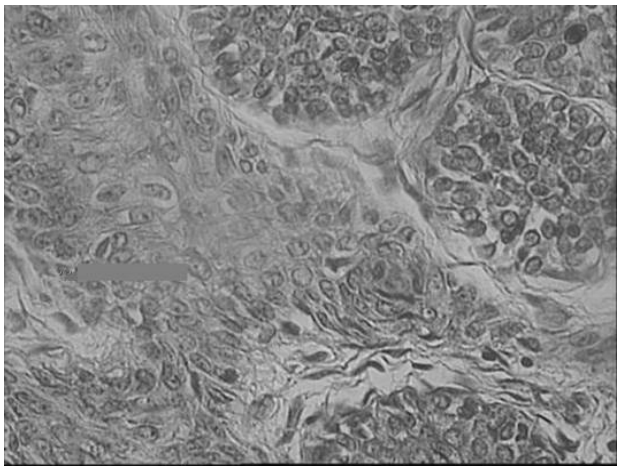


Figure 2C : Large and small nests with peripheral palisading, surrounded by stromal condensation.



Tumour cells are small and basophilic, with minimal cytoplasm but without pleomorphism (Figure 2D). A normal mitotic activity is noted confirming the benignity of histologically typical trichoblastoma. Surgical margins were normal. Successful graft was made with good vitality, luckily for the patient, who was widely satisfied and refused the expansive procedure proposed to achieve an optimal aesthetic result. There was no evidence of tumour recurrence after a 5 year-follow-up period.

Figure 2D : Basaloid and uniform tumoral cells without pleomorphism



DISCUSSION

Trichoblastomas are solitary, small and well-circumscribed lesions that occur in the deep dermis and subcutaneous tissue, on any hair follicle-bearing location but the preferred anatomical sites are the head (especially face and scalp), the neck, and more rarely trunk and proximal extremities. The perianal region also appears to be a site of predilection. They can arise at any age, but are more common in adults (fifth to seventh decades) with no sex predilection. There is no relationship with familial multiple trichoepitheliomas. The majority of trichoblastomas are less than 2 cm in size, but as in our case, giant trichoblastomas can sometimes be seen, reaching several centimetres (up to 10 cm) [1, 7]. They should be isolated because they may be confused with malignant neoplasm owing to their size and frequent post excision relapse. Clinically, Trichoblastomas are often skin-coloured and rarely ulcerated and present as nodular form (like for our case) or more rarely as infiltrative plaques [8].

Although giant solitary trichoblastoma shows histological features similar to the usual trichoblastoma, there are important differences.

Classical trichoblastomas are well-circumscribed, symmetrical dermal tumours, with no epidermal connection. The giant solitary variant characteristically involves the subcutaneous fat in addition to the dermis [1]. The tumour is characteristically biphasic, being composed of lobules of basaloid cells intimately associated with a conspicuous fibromyxoid stroma. Large lobules of basaloid cells are arranged in cords, strands and nests, with extensive anastomosing and mixing of patterns from one area to another, realizing massive or mosaic patterns. Peripheral palisading of cells in larger nests is often present. Mitotic activity varies from tumour to tumour, but is always present and can be brisk. Sometimes, tumour nests are colonized by dendritic melanocytes and can be pigmented [6]. Abrupt keratinisation within the nests, with keratin cyst formation, is less frequently seen than in conventional trichoblastoma. The fibromyxoid stroma is an important, integral component of the tumour. It is typically composed of plump stellate to fusiform spindle cell fibroblasts with eosinophilic cytoplasm. The ratio between the epithelial component and the stroma varies. The tumours with predominance of the epithelial component are sometimes referred to as "trichoblastic trichoblastomas" (like our case), whereas those with stromal predominance are often called trichoblastic fibromas [3- 5]. The stromal component resembles the perifollicular fibrous sheath and often shows focal aggregation of mesenchymal elements in proximity to the periphery of basaloid nests, recapitulating the papillary mesenchymal body. The evidence of trichogenic differentiation is present in the form of abortive hair papilla formation (so called papillary mesenchymal bodies), consisting in an indentation of the stroma into nests of epithelial cells, reminiscent of the relationship between primitive hair papillae and hair bulbs [4]. Trichoblastic epithelial components associated with stereotyped stroma, chiefly the follicular papilla, must be present to establish the diagnosis with surety. Trichoblastomas are also often characterized by a cleft between their mesenchymal component and the surrounding stroma. During surgery, the tumour typically can be shelled out from the adjacent tissue [2].

Trichoblastoma and basal cell carcinoma (BCC) have some overlapping clinical features. But, it is important to distinguish

between the two lesions because treatments and prognosis are completely different. The lack of epidermal origin, more conspicuous stroma with prominent papillary mesenchymal bodies and absence of retraction artefact between the tumour epithelium and stroma are useful diagnostic features in excluding basal cell carcinoma [9]. Furthermore, the analysis of typical genetic alterations of these tumours can provide a better understanding [2].

Trichoblastomas can occur sporadically as predominantly dermal neoplasms or arise as secondary tumours within nevus sebaceous. In fact, trichoblastoma is thought to be the most common neoplasm developing in nevus sebaceus, and it has often been formerly interpreted as basal cell carcinoma [10]. Our patient did not report any pre-existing lesion before the onset of the tumour.

Rare cases of trichoblastic carcinomas have been reported and should be differentiated from giant forms of trichoblastoma [9, 11, 12, 13].

They are characterized by a propensity for local recurrence with potential metastatic spread. While simple excision is curative in benign trichoblastoma (as in our patient) with a minimal margin (1cm in our case), carcinological excision with clear margins is recommended in trichoblastic carcinomas with long-term follow-up.

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