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CASE REPORT

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Typical MDCT Angiography Findings of an Unusual Cutaneous Neoplasia; Masson Tumor

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Summary

Background: Intravascular papillary endothelial hyperplasia (IPEH), also known as masson tumour, is a lesion composed of proliferating endothelial cells.

Case Report: In this article we explained clinical, histological and radiological features of IPEH involving the scalp, localized on the left side of the skull and in the periauricular region.

Conclusions: Radiologically, intravascular papillary endothelial hyperplasia could be misdiagnosed as malignant or benign vascular tumour. On cross-sectional imaging it is useful demonstrating the extremely vascular component of IPEH. But IPEH has no specific radiologic features that we can use to differentiate from the aforementioned lesions. Due to that, histopathological examinations are needed to diagnose IPEH.

MeSH Keywords: Angiography • Hyperplasia • Multidetector Computed Tomography

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Background

Intravascular papillary neoplasia also known as Masson tumor was first described by French pathologist Pierre Masson in 1923 [1]. IPEH is now believed to be a benign intravascular process that can take place during thrombus organization [2]. IPEHs comprise approximately 2% of vascular tumors of the skin and subcutaneous tissue. In this article we tried to explain clinical, histological and radiological features of IPEH involving the scalp, located on the left side of the skull and in the periauricular region.

Case Report

A 32-year-old female patient presented at the dermatology clinic of our hospital. Her complaint was a palpable mass near her left ear, which had been growing rapidly for 1 month. She did not have any comorbidity or dermatological problems. She also mentioned that the mass was too tender to touch. Our physical examinations within the left retropineal area showed a soft painful mass, approx. 5 cm in size, surrounding the left ear. The patient was examined with CT (GE-optima660, 64-slice multi detector CT) and a bolus tracking method with a biphasic angiography protocol. During examination, non-ionic contrast agent (70 mL of iopamidol 300/100) was given to the patient at a rate of 4 mL/min. After contrast administration, 20 mL of serum was given to the patient at the same rate. On the following day, to determine the inner nature of the lesion, a 3 Tesla MRI (Siemens Magnetom Skyra) examination was carried out without contrast enhancement. The following images were obtained to examine the lesion: T1 axial, T2 axial, SWI axial. Our CT examinations showed that the mass lesion did not have any calcifications. Moreover, we found smaller multiple soft-tissue masses spreading on the left side of the scalp (Figure 1). On dynamic contrast-enhanced CT examination, those lesions showed peripheral and asymmetric contrast enhancement. When measuring the density of the lesion, we found that it had the same density as arteries in the arterial phase and the same density as

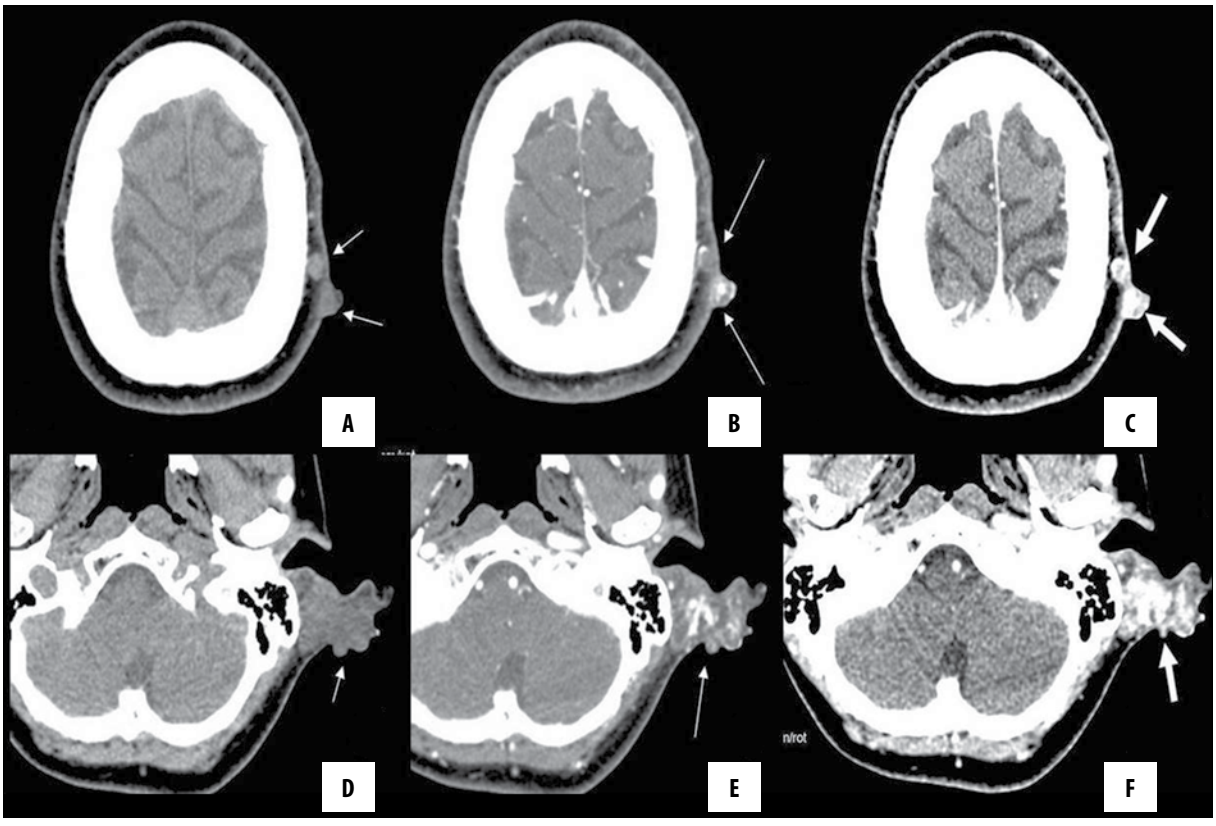

Figure 1. On dynamic CT examination, axial sections showed a characteristic contrast retention in the nodular lesions on the scalp. On the sections near vertex (**A–C**) and through the left ear (**D–F**) the lesions are hypodense in pre-contrast images (short arrows), there is central or eccentric contrast retention in the arterial series (long arrows) and heterogeneous hyperdensity in the venous series (thick arrows).

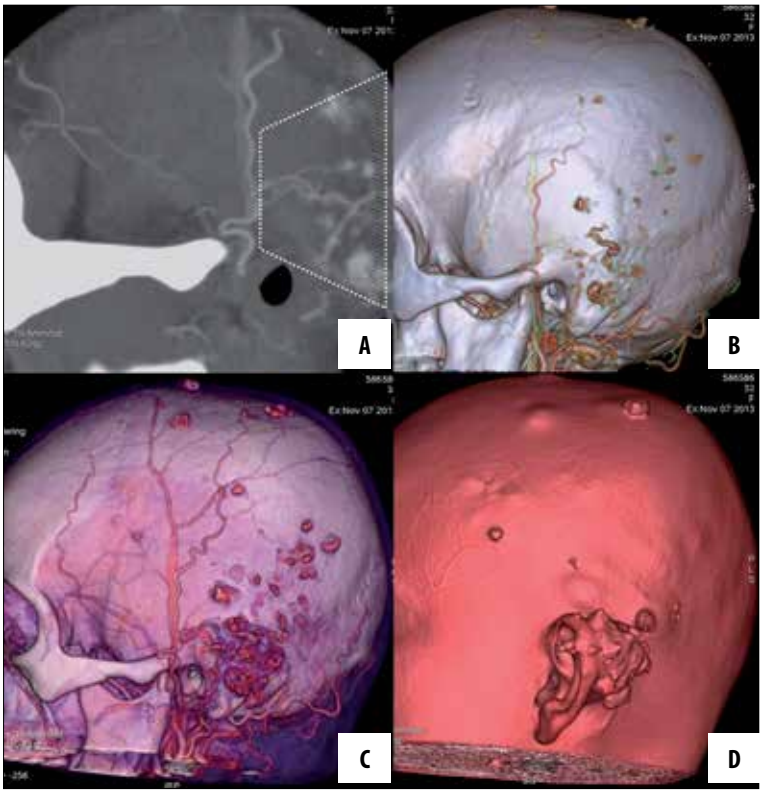

Figure 2. The demonstrative features revealed on reformatted and 3D CT images. A sagittal maximum intensity projection (MIP) reformatted image (**A**) revealed patchy enhancement at in the venous drainage areas (dashed-line area) around the concomitant arteries. On the series with an increased contrast dose (**B–D**), the location and distribution of soft tissue masses became clearer.

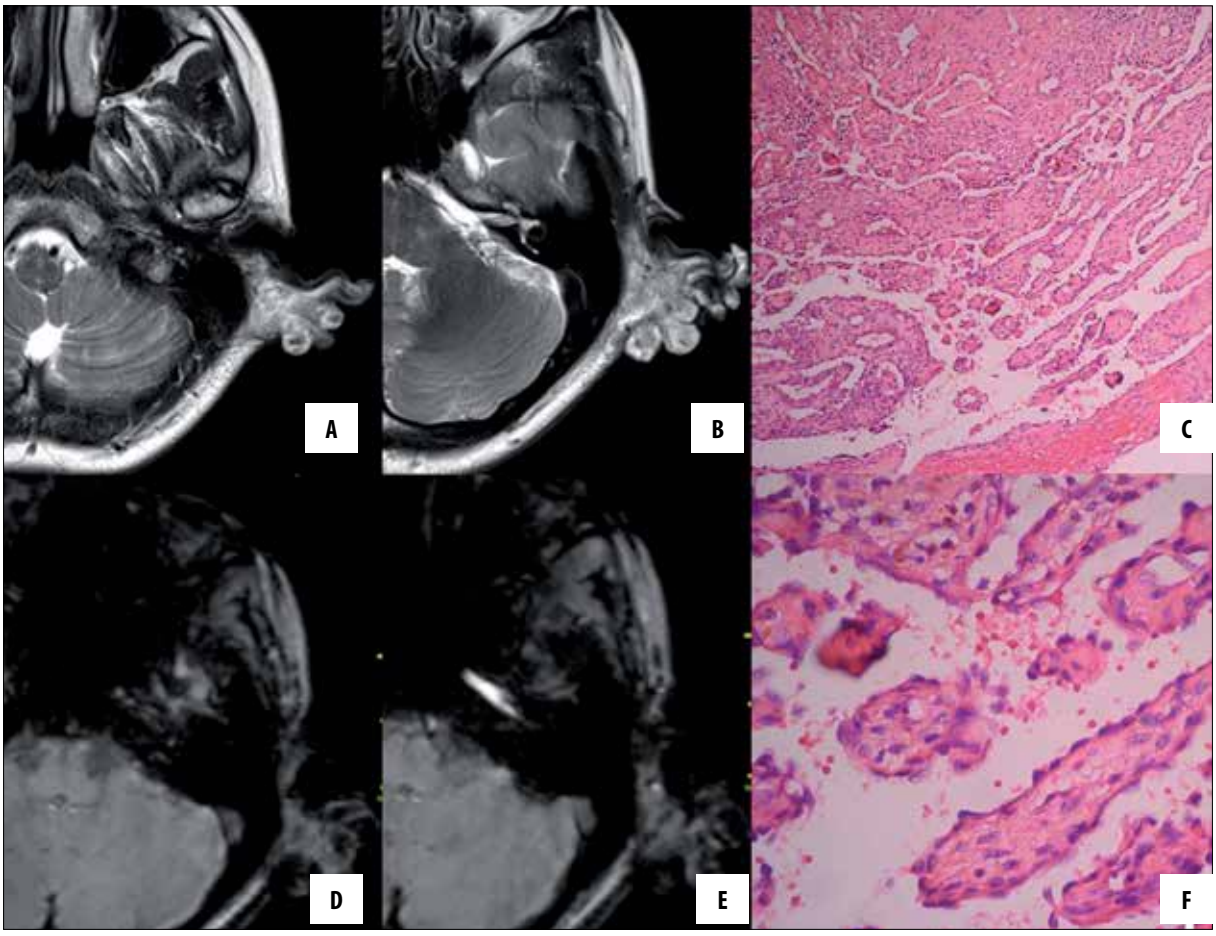

Figure 3. In the superior, axial T2W MRI images (**A, B**) through the skull base and auricles, the lesion is isointense with respect to the scalp, there is no evidence of invasive or infiltrative growth pattern, and hypointense deposition due to slow flow. Signal loss in this lesion on SWI images (**C, D**) shows an intensely hypervascular nature of the tumor. The histologic sections (**E, F**) (H&E, **E**; ×100, **F**; ×400) revealed some vague papillary proliferations of epithelioid cells with fibrinous materials. The peripheral portions show fibroblastic proliferations with some metaplastic formation. In high-power view in f, there are vague papillary configurations of epitheloid endothelial cells with occasional distinctive nucleoli and vesicular nuclei.

veins in the venous phase (Figure 2). On T2w MRI images, the lesion was hyperintense when compared with the scalp and did not cause any lytic reactions within the external auditory canal or the cartilaginous tissue of the left ear. Moreover, there was no evidence of perilesional reactive changes (Figure 3). Afterwards the patient underwent excisional biopsy. The biopsy material revealed histopathologically a Masson's tumour. The most important microscopic features of the specimen included papillary proliferations of epithelioid cells that were composed of hemorrhage and fibrinous thrombus. Another significant feature was irregular nuclei of cells due to high proliferation rate. After diagnosis, the patient was subjected to surgery and successfully treated.

Discussion

In 1923, IPEH was described as a "neoplastic lesion of endothelium that usually occurs in the head region" by Masson, a French pathologist. Nowadays, it is considered as a proliferation of endothelium, caused by extraordinary thrombus formation due to stasis of blood in vessels [3]. Local hemodynamics is also considered responsible for this tumor, e.g. growth of endothelium [4]. Three different types of IPEH have been reported. The primary form occurs in distended vessels and the secondary form occurs as a consequence of a previous vessel pathology like hemangioma, pyrogenic granuloma or varicose veins. The third one is an undetermined form, belonging to neither of the first two categories, and is of extravascular origin [5]. Although our case showed a rapid progression, IPEH is usually a slow-growing, painful nodule, that can show a wide variation in age at onset (1–70 years) and female predominance. In surgery, IPEH tends to bleed much and can look the same as an organized hematoma. Curative treatment of the Masson's tumor is complete surgical resection. IPEH most commonly occurs in the skin and subcutaneous soft tissue [6]. Unfortunately, IPEH does not have any specific radiologic features. Radiologically it can be misdiagnosed as hemangioma, angiosarcoma or metastasis of kidney tumours. The final diagnosis has to be made based on histopathological examination.

In our case we found that contrast fixation patterns of the Masson's tumor were the same as arteries in the arterial phase and veins in the venous phase. Despite that

aggressive contrast enhancement, there was no evidence of infiltration of soft tissues or skull but it spread with satellite masses. These findings may be helpful for differential diagnosis of the Masson's tumor from other lobulated contour hyper-vascular soft tissue masses, which usually show an infiltrative pattern. IPEH of the scalp is a rare benign vascular tumor although it is rapidly progressive and appears to be malignant. Due to its rarity, it is radiologically misdiagnosed. Thus, final diagnosis requires a careful histological examination. These lesions may grow rapidly and reach an enormous size, and generally recur if incompletely resected. In a patient with a a hyper-vascular subcutaneous lesion, IPEH should be remembered of in the differential diagnosis. Conversely to the literature, we believe that CT examinations must be obligatory in a patient with a suspected Masson's tumor, for detection of IPEH lesions in case of which complete surgical resection is curative. Moreover, imaging features of that tumor are much more characteristic in CT than MRI.

Conclusions

Radiologically, intravascular papillary endothelial hyperplasia could be misdiagnosed as a malign or benign vascular tumour. On cross-sectional imaging, it is useful to demonstrate the extremely vascular component of IPEH. Measuring the density of the lesion in CT and comparing the obtained values with arteries and veins showed that it has the same density values as arteries in the arterial phase and veins in the venous phase, which can be very useful for diagnosis. However, all in all, IPEH has no specific radiologic features that could be used to differentiate it from the above mentioned lesions. Due to that, histopathological examinations are needed for diagnosis of IPEH. The treatment of İPEH is complete surgical resection. Long-term follow-up of some large case series did not show any metastases [7]. However, there are some cases of recurrence in the literature, in which chemotherapy was used as adjuvant therapy, but the efficacy of chemotherapy is not clear yet [8].

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