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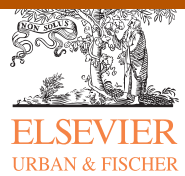

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Intravascular papillary endothelial hyperplasia at the origin of internal carotid artery: A rare cause of stroke

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KEYWORDS
Intravascular papillary endothelial hyperplasia;
IPEH;
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Summary Intravascular papillary endothelial hyperplasia (IPEH), also known as Masson's tumor, is a rare, generally considered a non neoplastic vascular lesion, caused by an abnormal endovascular proliferation of endothelial cells.

We describe, as far as we know, the first case of this lesion, localized at the origin of the internal carotid artery, which was responsible for an ischemic stroke. Although this entity is very rare, it is important for the clinician to become familiar with this lesion, since the complete removal of the lesion is the only treatment of choice. A partial removal may lead to further clinical events.

Introduction

Intravascular papillary endothelial hyperplasia (IPEH) is a relatively uncommon benign and non-neoplastic vascular lesion [1—4]. Firstly described by Masson in 1923, as an endothelial proliferation associated with thrombosis and fibrin deposition, leading to obliteration of the vascular lumen [1—4]. Histologically it is characterized by the presence of endothelium-lined papillary structures composed by a single layer of plump cells around a fibrin core that sometimes forms irregular anatomizing clefts, simulating an angiosarcoma [5—8]. However, the absence of cellular polymorphism, mitotic activity and necrosis represent a differential feature of IPEH [5]. The prognosis of this lesion is excellent, and recurrence is an unusual finding. It is cured by simple excision in primary forms, and by treating the underlying condition in the secondary ones [6,8]. Its pathogenesis is believed to be associated with trauma, but it has also been reported as an unusual form of organized thrombus [6—9].

Case report

A 43 years old female with neither previous history of neurological diseases nor vascular risk factors other

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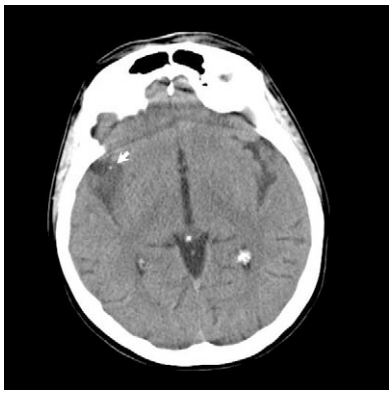

Figure 1 Brain CT: the ''dot sign'' (arrow) is visible in a M_2 branch of the right middle cerebral artery.

than smoking custom, was admitted to our Neurological Department — Stroke Unit, because of a left hemiparesis. She had felt her left arm somehow weak, strangely ''cold'', in the previous afternoon, but believing that this was related to fatigue she went to sleep. On admission a mild left sided sensorimotor hemiparesis was found with a NIHSS of 8.

The brain CT scan performed at the admission showed a ''dot sign'' in a M_2 branch of the right Middle Cerebral Artery (MCA) (Fig. 1) and a right fronto-parietal ischemic stroke (PACI) (Fig. 2). The EC US revealed at the origin of the internal carotid artery, an hyperechogenic lesion (Figs. S1 and S2). It was somehow different from an atherosclerotic plaque and more similar to a soft tissue mass. Its echogenicity was homogenous and, in its distal portion, it was partially separated from the arterial wall, but no flapping movement was evident (video). The lesion occupied more than 75% of the cross sectional area of the vessel, but no increased velocity was present (Figs. S3, and 3). At TCCD all the major intracranial arteries were insonated; an asymmetry of the MCA velocities (R < L), with a Zanette index of 26.39 suspicious for a right MCA distal occlusion, was found (Figs. S4 and S5). The Angio-CT confirmed these features (Figs. 4 and 5). The patient was evaluated for vascular risk factors, dietary

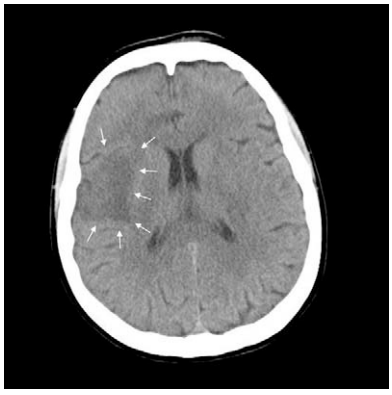

Figure 2 Brain CT: the right sided hypodense ischemic lesion is clearly detectable (arrows).

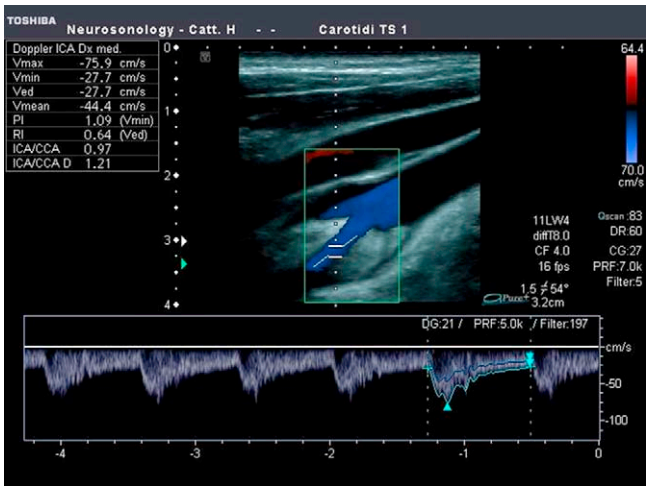

Figure 3 Extracranial carotid ultrsound examination. Right internal carotid artery and the flow waveform.

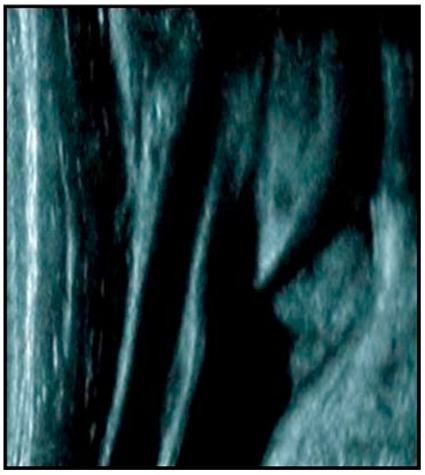

Figure 4 Internal carotid artery by extracranial carotid ultrsound examination.

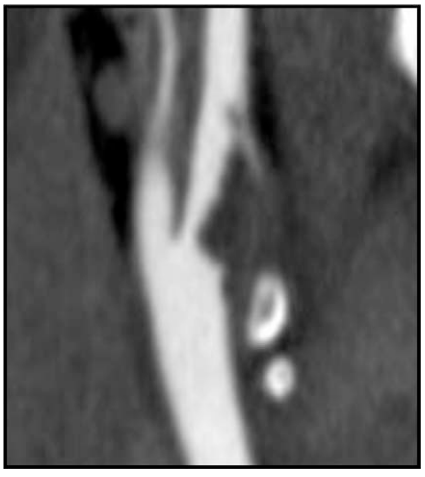

Figure 5 Internal carotid artery by Angio-CT performed the same day of ECUS.

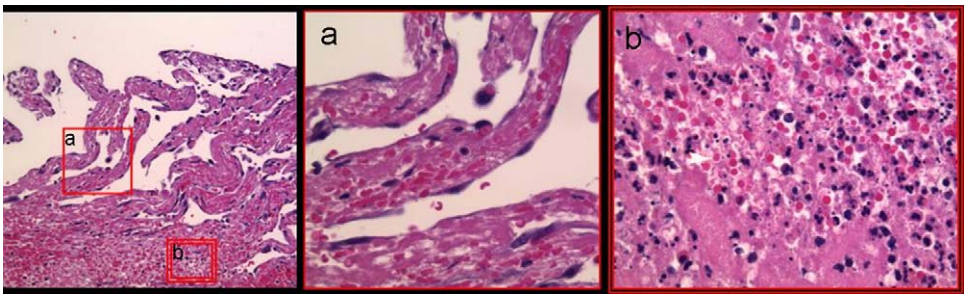

Figure 6 Histological specimens (EE stain) at different magnifications (a and b) showing an endothelial hyperplasia with a papillary distribution, surrounding a fibrin core with leucocytes and erythrocytes.

factors (Folate, B12), and methylene tetra hydro folate reductase (MTHFR) polymorphism; a thrombophilic screening was also performed. The condition of homozygosis for MTHFR was present. The histological diagnosis was consistent with a diagnosis of IPEH (Fig. 6): a marginal endothelial papillary hyperplasia, surrounding a fibrous and hematic material like an organized thrombotic formation, was described.

From the therapeutical point of view an antiplatelet therapy was started at the admission and the following days the clinical condition progressively improved. The vascular surgeon was then consulted and a surgical procedure was performed to remove the lesion.

When dismissed the patient was asymptomatic, the NIHSS equal to 0 and she did not suffer from any other symptom during the following 2 years. The EC US (Figs. S6 and S7), 2 years later, revealed some hyperplasia at the origin of the ICA possibly representing the over-expression of the post-CEA neoendothelial growth or the evolution of the incompletely removed lesion and showed that a longer follow-up is necessary.

Discussion

This pathology is quite rare compared with the common atherosclerotic lesions located at the carotid bifurcation; nevertheless the ultrasonographer may suspect it if no other atherosclerotic wall modifications are found elsewhere at the extracranial level, when the lesion is partly separated from the arterial wall in its distal portion and when no flow modification is found despite the huge lesion's dimensions. Moreover it is relevant to make the diagnosis for the clinician, since this lesion is highly prone to induce thrombus formation on its surface, with the possibility of embolic events. Early CEA is recommended and it is again relevant for the surgeon to suspect this diagnosis since, if the lesion is not completely removed, it can grow back again, with the risk of further embolic events.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.permed.2012.04.006.

References

[1] De Courten A, Küffer R, Samson J, Lombardi T. Intravascular papillary endothelial hyperplasia of the mouth: report of six cases and literature review. Oral Diseases 1999;5:175—8.
[2] Matsuzaka K, Koike Y, Yakushiji T, Shimono M, Inoue T. Intravascular papillary endothelial hyperplasia arising from the upper lip. Bulletin of Tokyo Dental College 2003;44:55—9.
[3] Devi M, Nalin Kumar S, Ranganathan K, Saraswathi TR. Oral intra vascular papillary endothelial hyperplasia in the floor of the mouth. Indian Journal of Dental Research 2004;15:149—51.
[4] Wang XY, Namiq A, Fan F. A 55-year-old woman with a buccal mass intravascular papillary endothelial hyperplasia. Archives of Pathology and Laboratory Medicine 2006;130:877—8.
[5] Inalöz HS, Patel G, Knight AG. Recurrent intravascular papillary endothelial hyperplasia developing from a pyogenic granuloma. Journal of the European Academy of Dermatology and Venereology 2001;15:156—8.
[6] Kim D, Israel H, Friedman M, Kuhel W, Langevin CJ, Plansky T. Intravascular papillary endothelial hyperplasia manifesting as a submandibular mass: an unusual presentation in an uncommon location. Journal of Oral and Maxillofacial Surgery 2007;65:786—90.
[7] Santonja C, De Sus J, Moragón M. Extramedullary hematopoiesis within endothelial papillary hyperplasia (Masson's pseudoangiosarcoma) of the tongue. Medicina Oral Patologia Oral y Cirugia Bucal 2007;12:E556—9.
[8] Anthony SG, Mudgal CS, DeLaney TF, Shin RD, Raskin KA, Ring DC. Recurrent intravascular papillary endothelial hyperplasia of the right middle finger treated with radiation therapy. Journal of Bone and Joint Surgery (British Volume) 2008;90:95—7.
[9] Cagli S, Oktar n, Dalbasti T, Islekel S, Demirtas E, Ozdamar N. Intravascular papillary endothelial hyperplasia of the cental nervous system. Neurologia Medico-Chirurgica 2004;44:302—10.