Chronic spontaneous diploic hematoma

Case report

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The authors report the case of a 23-year-old woman with café-au-lait spots and axillary and inguinal freckling who presented with a diploic chronic spontaneous hematoma of the left parietal bone. To the authors’ knowledge, this case represents the first description of a diploic hematoma in a patient with stigmata of neurofibromatosis Type 1 unrelated to head trauma. Plain skull radiography showed an osteolytic lesion with well-circumscribed margins, corresponding to the hematoma, together with exuberant perilesional vascular markings. Angiography demonstrated an incidental aneurysm of the left suprachiasmatic internal carotid artery and an unusual cortical venous drainage toward the diploic vessels. The blood flow of these vessels on the right hemicranium was sluggish and exhibited enlarged diploic venous lacunas. The authors hypothesize that the hematoma was formed by both an abnormal venous drainage toward the diploic vascular net, together with a vasculopathy that caused stenosis and obstruction of the normal drainage pathways from these vessels. (DOI: 10.3171/2010.12.JNS101589)

Key Words • spontaneous diploic hematoma • intracranial aneurysm • neurofibromatosis Type 1 • Legius syndrome • skull vault neoplasm

Abbreviations used in the paper: AVF = arteriovenous fistula; NF = neurofibromatosis; NF1 = NF Type 1; NIH = National Institutes of Health.

This article contains some figures that are displayed in color online but in black and white in the print edition.
CDNA sequencing showed no nucleotide variation in the codifying region for the *NF1* gene.

**Operation.** We noted a slight protuberance on the external table of the bone, which appeared thinned and bluish in color (Fig. 3 left). We made several bur holes surrounding the lesion that led to profuse hemorrhage from the dilated diploic venous channels. On elevating the bone, ample cavities were observed within the lesion (Fig. 3 right) that contained an apparent organized hematoma. This mass extended within the cranial cavity and compressed the adherent dura mater without infiltrating it. Following excision of the bone lesion and the adjacent dura mater, we performed duraplasty and cranioplasty.

**Histological Examination.** Histopathologically we observed communicating diploic cavities that produced bone expansion and thinning of the outer table, which was only 1 mm thick on a surface of 5 × 3 cm. On microscopic inspection, there was marked erosion of the inner table with cavities filled up with orange-reddish coagulated hematic material. Both the cavities’ content and the extraosseous tumor corresponded to densely organized fibrinous-hematic material with fibrous-hyaline peripheral zones, within which there was no viable cellularity (Fig. 4). We observed focal areas of hemosiderophagocyte and plasmatic cells. No vascular structures were detected with optic or special techniques. Fusiform or multinucleated cells that would suggest a neoplastic nature were not identified. The final histopathological diagnosis was organized hematoma with no vascular or neoplastic components.

**Postoperative Course.** Postoperative CT scanning (Fig. 1E) revealed a thickened diploe with abundant dilated venous channels in the unoperated right skull. Four-vessel angiography that included the external carotid arteries showed an incidental left supraclinoid carotid artery aneurysm (Fig. 1F), together with unusual bilateral venous drainage toward the diploe (Figs. 5 and 6). In the right frontal region, there was cerebral drainage...
toward the diploic vascular net that diverted the intraosseous blood flow to the frontal and parietal regions, which exhibited an ectatic behavior without draining into any intracranial venous sinus or extracranial veins (Fig. 5). On the left side, there also was an anomalous drainage in a diploic vein, with an ample course, that exited the skull to join the posterior cervical net (Fig. 6). No contrast flow toward the diploic vascular network, originating from the external carotid arteries, was observed.

The carotid artery aneurysm was successfully embolized. The patient’s subsequent course was satisfactory during a 3-year follow-up period.

Discussion

Intradiploic Hematoma and Head Trauma

We found only 4 reports of histopathologically verified organized diploic hematoma causing skull vault erosion unrelated to a preexisting pathological condition.\(^3,4,18,21\) Except for the case reported by Yücesoy and colleagues\(^21\) of a 25-day-old boy born after a complicated labor, all authors stressed the existence of a mild head injury occurring several years before detecting the presence of the intraosseous hematoma. Our patient did not recall any previous traumatic event, and, in addition, we consider it would be difficult to relate the presence of an organized hematoma of recent onset with the occurrence of a remote mild head injury, thus indicating the obscure origin of the hematoma in these cases.

Traumatic osseous cysts, giant cell granuloma, or aneurysmal bone cyst are osteolytic lesions of uncertain origin that have been associated with possible posttraumatic bone hemorrhage, but none of the histopathological characteristics was present in our patient’s lesion.

Diploic Hematoma From Intraosseous Vascular Malformation or Tumor

There have been infrequent reports of skull osteolytic lesions due to diverse tumors associated with hemorrhages within the adjacent epidural space, but in all these instances the existence of the lesion was obvious after histopathological examination. There have also been occasional reports of diploic cavernous hemangioma with significant intraosseous, epidural, or subdural hemorrhage, but similarly the nature of the underlying lesion was evident by microscopic study.

Although the organized hematoma in the present case seemed to be of relatively recent development, the appearance of the mass with bone thickening, thinning of the outer cortical layer, and the irregular sclerotic bone margins denote a slow and chronic process with an expanding force that caused erosion of the internal table. There exists the possibility that a benign and slow-growing tumor, as in some of the aforementioned conditions, might have bled and resulted in total destruction of the lesion, although we think it would be highly improbable not to have found some vestige of the primary cause in the histopathological study corresponding to the en bloc excised tissue.

Infrequent cases of spontaneous AVFs of the diploe,
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with enlarged diploic venous channels, have also been described. In our case, there was the possibility of a diploic AVF that expanded the bone, either by vascular pulsations or by the expansion of the veins belonging to the AVF, even perhaps by repeated bleeding, which finally would destroy the AVF itself, thus explaining the absence of AVF remnants on the histopathological examination.

Nevertheless, the findings of dilated diploic vessels and lacunas on the right side of the skull are not justified by a diploic fistula and suggest the existence of a more widespread vascular anomaly, which would justify the bone erosion and hemorrhages on the left side at the time that it would mean an incipient stage of the anomaly on the right hemicranium.

**Fig. 5.** Right internal carotid artery angiograms in the venous phases. **Left:** Anteroposterior image showing blood drainage toward the diploic veins (black arrows) originating from frontal cortical veins. A cortical vein that does not drain into the sagittal sinus (white arrows) is clearly visible; rather, this vein drains into the diploic vessels. **Right:** Lateral image demonstrating ectasia of the diploic vein (black arrows). The contrast persisted within this vessel even when it had been completely washed out from the intracranial venous sinuses (image not shown). Note that the diploic vein does not drain in any of the intra- or extracranial venous networks.

**Fig. 6.** Postoperative left carotid artery angiograms in the venous phases. **Left:** Anteroposterior image revealing venous drainage toward the diploic veins (black arrows). **Right:** Oblique image showing a large intraosseous trajectory of the diploic vein (black arrows) that ends draining into the posterior cervical vascular network.
Anomalous Venous Drainage, NF1 Vasculopathy, and Legius Syndrome

The patient we report on presented with a carotid artery aneurysm and bilateral cerebral venous anomaly with cortical veins that drained to the diploic veins rather than the intracranial venous sinuses (Figs. 5 and 6). On the unoperated right side, the diploic lacunas and channels were enlarged (Fig. 1E), and there was an obvious ectasia of the blood flow that finally stopped without showing an evident drainage (Fig. 5 right). In our view, this feature is difficult to explain. However, the café-au-lait spots and widespread axillary and inguinal freckling that the patient exhibited evoke the cutaneous marks of von Recklinghausen disease despite the fact that the patient did not completely fulfill the NIH clinical criteria for the diagnosis of adult NF1. Cases that do not satisfy the recognized clinical or genetic criteria, however, may constitute the need to establish additional forms of NF1, according to the NIH. Recent reports indicate that patients with café-au-lait spots with or without freckling, Lisch nodules, neurofibromas, or other characteristic lesions of NF1 may have mutations in the SPRED1 gene of the long arm of chromosome 15 (15q13.2) known as Legius syndrome (sporadic cases of frequent occurrence). Messiaen et al. reported on 5 of 15 patients older than 20 years who were carriers of SPRED1 loss-of-function mutation and who did not present with the number or size of café-au-lait spots, as the first criterion of the NIH requires, for a diagnosis of NF1. Accordingly, we cannot rule out that our patient was not a carrier of a mutation in one of the genes that impair protein coding within the Ras-MAPK signaling pathway, as has been linked to the development of NF1 vasculopathy.

Knowledge of NF1 vasculopathy has grown in the last decades, but its incidence is unknown, as many patients remain asymptomatic their entire lives. The significance of this entity, in all likelihood, remains poorly recognized. The incidence of vasculopathy in the recently defined Legius syndrome and in other NF-like syndromes is a matter that deserves further investigation.

Vasculopathy in NF1 is characterized by cell accumulation on a vessel's intima leading to hyperplasia that narrows the vessel's lumen. The best known changes are those that affect the systemic arterial tree, producing stenosis, ectasia, occlusion, aneurysms, pseudoaneurysms, rupture of the blood vessels, or development of fistulas. In a review of 42 cases with cerebrovascular involvement culled from the literature, Sobata et al. found a higher prevalence in females, and a higher incidence of stenotic vascular lesions (71%) than aneurysms (19%) or a combination of both (10%). According to age, the incidence of aneurysms increased up to 35% in adults.

Vasculopathy in NF1 can also involve the veins, although this has received little attention in the literature. There have been isolated case reports of aneurysm, ectasia, and stenosis of the internal jugular vein, spontaneous rupture of the common iliac vein, superficial femoral vein dysplasia, tibialis posterior vein obstruction, retinal vein occlusion, “corkscrew” tortuosity of retinal veins, and optic disc venous anomalies.

Possibly, in our patient, the anomalous cerebral venous drainage and the obstruction of these diploic veins might constitute 2 different events, but their coincidence in the same vessels caused hypertension in the diploic veins, leading to venous enlargement and, finally, to bone erosion and epidural bleeding. The origin of the stenosis of the diploic vessels is both intriguing and unknown. Our hypothesis that the vasculopathy due to impaired Ras-MAPK signaling pathway is responsible for the diploic vascular ectasia is speculative; it is based only on the presence of a carotid artery aneurysm in a 23-year-old woman with café-au-lait spots and axillary and inguinal freckling.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: González-Tortosa. Acquisition of data: Martínez-Lage, Ferri-Núñez, Parrilla. Drafting the article: González-Tortosa and Martínez-Lage. Critically revising the article: González-Tortosa, Martínez-Lage. Reviewed the article: González-Tortosa and Martínez-Lage. Reviewed the final version of the manuscript and approved it for submission: all authors. Study supervision: González-Tortosa.

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