Considerations on port-wine stains and their laser treatment

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Capillary-venous malformations on the forehead with intracerebral connections: sinus pericranii.

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Introduction

Sinus pericranii was described in 1885 by Mastin as a venous blood tumor of the vault of the cranium communicating with the intracranial venous circulation, especially through the medium of the superior sagittal sinus. Mastin collected several descriptions from authors in the German and French literature starting in 1826, who mentioned similar findings (1).

The diagnosis of sinus pericranii is made when a combination of clinical and radiological features exists. A blue discoloration is present, sometimes with a swelling on the outside of the central forehead or the lateral side of the skull, which disappears under manual compression and aggravates during exercise, crying, coughing and head down position (2,3,4). Its appearance can be as a local varix or a considerable swelling sometimes with additional capillary malformation (port-wine stain). Patients can complain about discrete headaches or are seen in the neurosurgical department with convulsions (6,7). However, most patients with sinus pericranii are without symptoms. They present themselves with a cosmetic wish for removal of the blue color and/or swelling on their forehead.

We describe seven patients who presented themselves with a sinus pericranii at the plastic surgical office or in our working group for vascular anomalies during the last five years. This rare anomaly is less well known. Standard angiography will not visualize the perforators through the skull. Special instructions have to be given when a sinus pericranii is suspected. Therapeutic management can be easy in case the problem is properly delt with.

Case 1

A 6 weeks old baby girl was admitted with a blue discoloration of her forehead, extending from the bridge of the nose into the hair bearing area in a parasagittal direction. Pregnancy and birth were uneventful. A bony irregularity was palpable in the affected region. Crying and horizontal position of the head caused considerable swelling. No neurologic symptoms were diagnosed. The swelling was compressible and no souffle was heard. At the age of 6 months angiography showed an intracranial venous malformation, draining through the frontal bone to a superficially located huge vein. A secondary outflow of the venous malformation was to the medial cerebral vein. CT-scan did not show a bony defect. No intervention was planned because of fear of a cerebral frontal infarct when the superficial vein would be occluded. At the age of 8 years she came back because of increased swelling of her forehead. Her development was normal. She had no complaints about her forehead except a cosmetic problem.

A new angiography was performed through the internal and external carotid artery. Visualization of vessels was performed in a head down position, with and without compression of the malformation. The malformation and transosseous connections were as well visualized through direct puncture venography. Only with compression of the malformation a normal drainage to the superficial sagittal sinus could be shown. An additional brain scan (Spect) was performed with and without compression of the malformation. Perfusion of the frontal lobes was identical in both situations.
Through a sagittal incision of the forehead the dilated veins were visualized. The affected skin was simultaneously excised. Three passages through the frontal bone were identified, two perforating veins were ligated, a third teared at the transition to the frontal bone, where the hole in the diploe was recorded on plain X-ray. All perforating holes were filled with bone wax. The outcome was uneventful with a good cosmetic result. The girl could be discharged two days after the operation and is well now, half a year later.

**Case 2**

This boy was first seen at the age of 6 years with a blue swelling on his forehead, present since birth, and growing commensurate with the child. An attempt to remove the swelling elsewhere ended in massive blood loss without removing the malformation. He had no complaints, however his parents requested removal because of cosmetic reasons. Internal carotid angiography showed an intracerebral venous malformation (Fig 1). At first through the arterial route no venous malformation on the forehead could be visualized under hypotensive general anesthesia. Again, head down position and normotension showed a connection between the transverse sinus to the superficial system on the forehead. The intracerebral venous outflow was normal. CT scan showed no bony defects. During operation the majority of the affected skin could be removed as well as the dilated veins. Perforators were ligated and bony defects were filled with bone wax. Healing was uneventful. The edge of affected skin still looks like a blue ribbon on the forehead, which he wants to have removed before going to high school. No swelling of the forehead was seen after the operation, with a follow up of 2 years.

**Case 3**

A 4-years old boy was seen with a red/blue discoloration of his nose and forehead, present since birth. No trauma or infection was known. Development was uneventful. The impression was one of a capillary malformation of the nose. No pulsations were felt or heard. Above his right eye dilated veins were visible. Angiography showed a capillary malformation of nose and left maxillary sinus. Intracranially, a very broad, pathologic venous sinus was seen at the skull base. The dilated vein on his forehead drained an intracranial venous anomaly in the roof of the orbit and lamina cribrosa (Fig 3). To our opinion, embolisation had a risk of intracerebral damage and would almost certainly result in a recurrence. No therapeutic intervention was undertaken. This patient is unavailable for follow up.

**Case 4**

A 13 year old boy had since birth a blue discoloration of the left side of the skull, extending in the lateral side of his upper eyelid and the sclera of his left eye (Fig. 2a). During childhood the affected area showed increasing swelling during strenuous activities. The malformation bled several times during the last year before presentation in our hospital. Phleboliths could be palpated in the affected area. The boy was checked
neurologically because of headaches. Physical examination, electroencefalogram, CT of the skull did not show any intracerebral defect. The extracranial swelling seen on contrast-enhanced CT scan (Fig. 2d) was confirmed with angiography. This investigation showed the venous outflow going from the intracranial to the extracranial veins (Fig 2b,c).

To prevent bleeding events of his eye, the patient was operated on his left eye, eyelid and left temporal region by an ophthalmologist and a plastic surgeon in a single session. During the same session some vessel mass in his ptotic upper eyelid was surgically removed. The vascular mass in his left temporal area was treated with a Nd-Yag laser, initially with good success. Slowly recurrent swelling on the forehead was noticed after 5 years.

Case 5

An 8 year old girl with a history similar to case two. She has been operated without problems with a follow-up of 4 years. On and around her nose and forehead telangiectasia remained which will be treated with the flash-lamp pulsed-dye laser.

Case 6

A 28 years old female had a similar history and appearance as case two. She was known with a blue discoloration on the forehead since birth. There were never health problems. The woman only wanted to know what the cause of the discoloration was and if this abnormality contained health risks for her. Because she decided not to have any treatment, no further investigations were performed. She felt relieved to hear a proper diagnosis and explanation of the anomaly.

Case 7

An 8 years old girl who was seen with an blue swelling on the bridge of her nose, extending to the right orbit, present since birth. Angiography showed distension of the ophthalmic vein developing after sinus thrombosis. Redistribution of flow through the veins and drainage of the cerebral vessels through the ophthalmic vein, were responsible for the extracerebral blue swelling. No therapeutic intervention was planned.

Pathologic examination of tissue removed from the operated patients (case 1,2,4,5) showed normal endothelial lining of the vessels.
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Legends to

Fig. 1a: Typical aspect of sinus pericranii with swelling on forehead and capillary malformation on right side of swelling.

Fig. 1b: Angiography: developmental venous anomaly intra cerebral of the same patient.

Fig. 2a: Lateral sinus pericranii extending from left eyelid and left parietal region to the vertex.

Fig. 2b: Shows angiographical findings with sinus on vertex. Inset: Location of venous drainage.

Fig. 2c: CT scan with extracranial swelling with phlebolith.
Discussion

We report on seven patients with a sinus pericranii who presented themselves with a cosmetic wish for removal. They were all seen in a period of five years in one hospital. Since we are a referral center for vascular malformations, this may account for the high incidence. More patients were seen than expected, in view of the sporadically published case histories. In five patients an additional intracerebral vascular malformation was seen. An extracranial operation was performed successfully in four patients.

Etiology

Both a congenital and a traumatic origin of the pericranial sinus have been proposed (8). Foetal injury during labor can be classified as trauma of the head. However, since several congenital cases are diagnosed in combination with congenital intracerebral malformations a pure traumatic origin seems unlikely (9). All our cases were considered to be congenital, except case 7.

Although most facial vascular malformations are not associated with intracerebral developmental anomalies, some authors mention relations to intracerebral malformations (6,8). In cases 1 and 2 in our series distended veins were associated with intra cerebral developmental anomalies. Case 3 presented with a capillary malformation of nose and maxillary sinus. Other associations with genetic implications are described: sinus pericranii in a Crouzon patient, sinus pericranii in another craniosynostosis patient, sinus pericranii in combination with blue-rubber bleb nevus syndrome or sinus pericranii with systemic angiomas (10,11,12,13,14).

Diagnosis

Analysis of the localization of the affected sites shows that the frontal region is most frequently involved (13). This is also the case in our patient series. When a midline frontal venous malformation is diagnosed and treatment is considered, an internal carotid angiographic evaluation must be performed. Through this procedure can be determined whether there is a pathologic communication between vascular malformation and intracerebral vessels. When an abnormal communication exists, it is of vital importance to find out whether this sinus drains normal brain and if drainage is guaranteed when these communicating veins are obliterated. Angiography is certainly advised when there is an association between an extracranial vascular swelling in combination with neurological symptoms (6,7).

All our patients were without neurological symptoms, although one boy (case 4) complained of headaches. This confirms data from others (13).

It is not always easy to visualize the vascular communication between the extracranial region and the underlying dural sinus during carotid angiography. X-rays taken in head down position or direct injection of contrast medium into the malformation make it possible to assert the diagnosis of sinus pericranii as we described in our first case. Kurosu et al. mentions a patient with hypoplastic veins (11). In case 2 normal venous outflow of the brain could only be visualized when the malformation was compressed externally. Without compression inadvertently a diagnosis of hypoplastic intracerebral
veins would have been made. It has to be brought under discussion whether Kurosu's patient really had hypoplastic veins or if the same phenomenon (case 2) led to a false diagnosis (11).
CT scan did not give additional information in our patient group. The diameter of the skull perforating holes, as confirmed in our patients during operation was 1-2 mm.

Therapy
Surgical removal is performed easily most of the time. All our cases were treated through a extracranial approach. Blood loss was minimal. In our series the operation in another hospital of one patient had to be stopped because of a tremendous blood loss, which is also mentioned by others. Careful preparation of the intervention seems mandatory. Packing of the sinus with bone wax was sufficient in our cases. Removal of the affected bone as suggested by Beers and Spector seems unnecessary to us (2,15).
Until now most reports on sinus pericranii are presented in neurosurgical or radiological journals. All our patients presented themselves at a plastic surgical unit or outpatient clinic for vascular anomalies.

References


