Melorheostosis — a very rare entity in neurosurgery

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Abstract

Melorheostosis is of minor importance in neurosurgery. A case of this disease is reported, which required neurosurgical activity and the literature is reviewed.

Keywords: Melorheostosis, intracranial space-occupying lesion, neurosurgical treatment.

1 Introduction

In 1922 LÉRI and JOANNY originally described a rare form of hyperostosis which is characterized by its typical cortical thickening of the bones [12]. Since the radiological appearance resembled melting wax dripping down a candle they called it flowing hyperostosis (hyperostose en culée) or melorheostosis from the Greek words melos for member and rhein for flow.

Initially the disease was thought to be monomelic, but now it is evident that it can include the limbs, skull, and trunk in one and the same individual. Although melorheostosis usually affects the long bones, it can involve any bone in the body [2]. Approximately 300 cases have been reported in the literature [20].

In most cases the patients complain of pain, stiffness, limitation of joint motion, or deformities. Sometimes the disease is accidentally discovered [9, 10, 11]. It tends to progress slowly in childhood and adult life [4, 17, 20]. Nowadays melorheostosis is considered to be a disorder not confined to bone, since the adjacent soft tissue is also often involved [5]. There are many reports on the association of melorheostosis with numerous mesenchymal abnormalities. These include extracranial arteriovenous malformations [8], linear scleroderma [4, 18], linear cutaneous vascular malformation [17], metaplastic cartilage formation within the mesenchymal tissue [13], congenital carpal tunnel syndrome due to such cartilage formation [1], and others. There is no gender predominance and no indication of a hereditary factor.

Aetiology remains unknown. It seems probable that the disease is caused by a primary defect of mesenchymal differentiation [14]. THOMPSON et al. stated that a primary mesoblastic defect arises in a part of a limb before its development into a definitive limb [18]. In other theories, nerval or vascular lesions, infections, or endocrine factors were considered to be the aetiological agent.

Due to the sclerotomal distribution of the affected bony and soft tissue structures it is proposed by MURRAY and McCREDIE that melorheostosis may be the result of segmental sensory nerve lesion [15]. KAWABATA et al. reported two cases of melorheostosis of the upper limb and considered the primary disorder to be related to the distribution of sensory nerves according to MURRAY and McCREDIE’s sclerotomal hypothesis [10]. MORRIS et al. suggested that the degenerative, inflammatory, and obliterative changes in blood vessels are the primary initiating factor [13].

Melorheostosis affecting the skull or spine is unusual [3, 7, 21]. A cranial or spinal form of melorheostosis requiring neurosurgical intervention is even more unusual. Only two cases
of spinal melorheostosis associated with neurological deficit due to intrathecal lipoma have been reported in the literature [6, 15].

2 Case report
To the best of our knowledge, we presented in 1987 the first case of melorheostosis with an intracerebral space-occupying lesion requiring neurosurgical treatment [19]:

A 13-year-old girl complained of unspecific pain in both legs. Laboratory values were normal. Radiographs of the tibia revealed the typical hyperostosis. Biopsy confirmed the diagnosis of melorheostosis. One year later she suffered from increasingly severe headache after a slight head injury. Plain films of the skull showed a giant sclerotic intracranial mass (Figure 1a + b). The CT scan demonstrated a calcified intracranial cauliflower-like space-occupying process (Figure 2). Since her general condition was good, the patient was not operated on because of the high risk of postoperative neurological sequelae. Only a transventricular biopsy was carried out. The histological examination yielded fibrous tissue with extensive calcification (Figure 3). No tumor cells were found. When the girl was 16 years old an incomplete occlusion of the foramen of Monro occurred and a shunting procedure was performed. Nevertheless, her condition got worse. Physical examination demonstrated weakness in both legs and balance problems. In addition, diencephalic disturbances were found. An attempt at total removal of the process failed. The patient died 15 days postoperatively due to pneumonia. The autopsy findings confirmed the melorheostosis (Figure 4) and revealed a meningioma of the cerebral falx adjacent to the calcified mass (Figure 5) as well as a neurinoma of the left acoustic nerve.

3 Discussion
The neurosurgeon has to deal with melorheostosis extremely rarely. Only three cases have been reported in the literature [5, 16, 19] in which not so much the bony changes but soft tissue abnormalities required neurosurgical intervention. GAUPP in 1949 reported a similar case of melorheostosis associated with meningioma in the fissure of Sylvii and neurinoma of the trigeminal and acoustic nerve [6]. He suspected that osseous and tumorous formation resulted from a common congenital failure during fetal development. WARZOK et al. pointed out the high frequency of tumors (particularly of the nervous system) in patients with melorheostosis [19].

![Figure 1a + b. Frontal and lateral radiograph of the skull showing the large intracranial calcified mass.](image-url)