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Myofibroma in the frontal region of the skull with a history of trauma in a 3.5-year-old child – a case report in the context of imaging diagnostics

Stefan Taront[A] ✉, Ewelina Mielewczyk[B], Ewelina Pilawa[C], Wojciech Poncyljusz[D]

Pomeranian Medical University in Szczecin, Department of Diagnostic Imaging and Interventional Radiology, Unii Lubelskiej 1, 71-252 Szczecin, Poland

[A] ORCID: 0009-0006-3132-2768; [B] ORCID: 0009-0003-7753-8028; [C] ORCID: 0009-0005-8500-0994; [D] ORCID: 0000-0002-5173-5635

✉ st.taront@gmail.com

ABSTRACT

Myofibromas in the orbit are considered very rare. According to literature data, 50% of myofibroma lesions occur in the neonatal period, and most of the remaining cases occur in children under 2 years of age. The diagnosis of myofibroma is possible based on histopathological examination. Morphological characteristics in imaging studies, patient's age, size of lesions and their number are helpful in making the diagnosis.

The presented case concerns a 3.5-year-old boy whose parents observed a rapidly growing tumor in the frontal region during a week and a half before the child was admitted to the hospital for further diagnostics. The interview revealed that the boy had suffered a head injury 2 months earlier, in the form of being hit by a swing. Since the injury, the frontal region has been under periodic surgical observation due to a suspected post-traumatic hematoma. The initial ultrasonud examination of the soft tissues of the frontal region revealed a poorly demarcated, non-movable, non-compressible mass with approximate dimensions of 3 x 5 x 5 cm. The lesion was filled with heterogeneous, hypoechoic content (solid or fluid); hyperechoic bands were visible within the lesion near the squamous part of frontal bone, which could correspond to calcifications or bone destruction. The Color Doppler examination did not clearly demonstrate vascularization within the lesion. The patient also underwent a magnetic resonance imaging (MRI) scan of the head, which revealed a tumor in the left frontal region measuring 56 x 39 x 52 mm, with intermediate signal intensity in T1- and T2-weighted images, which was contrast enhanced mainly in the peripheral parts of the lesion and did not show signs of restricted diffusion on diffusion-weighted imaging (DWI) sequence. Frontal craniotomy and left orbitotomy were performed. The tumor was removed from the anterior cranial fossa, left orbit and ethmoid. Fragments of bone infiltrated by the tumor were also removed and cranioplasty was performed. Myofibroma lesion was confirmed histopathologically. Apart from surgical treatment, during the entire treatment at the local clinic, the patient received 5 cycles of cytostatic treatment (vincristine) with good tolerance. The authors of this publication are inclined to assume that the injury that occurred in the boy in the described case could have been related to the later appearance of a myofibroma in the frontal and orbital regions. This case also emphases the value of control ultrasound examinations, which are the first and most easily accessible to assess the evolution of post-traumatic changes in paediatric patients.

Keywords: myofibroma; myofibromatosis; orbital neoplasm; trauma; diagnostic imaging; paediatrics.

INTRODUCTION

Myofibroma is a subtype of tumor called myopericitoma and usually arises in the dermis or subcutaneous tissue. Myofibromas can occur as solitary or multicentric lesions (myofibromatosis). Solitary myofibromas mostly arise in the skin and subcutaneous tissue of the extremities, head and neck, and trunk. In infants, myofibromatosis can occur in the viscera, including the liver and gastrointestinal tract, and in the brain and bones [1].

According to literature data, 50% of myofibroma lesions occur in the neonatal period, and most of the remaining cases occur in children under 2 years of age [2, 3]. Myofibromas in the orbit are considered very rare. In a publication analyzing 340 orbital tumors in children over a period of 60 years, only 1 case of myofibroma was described [4, 5].

It has been shown that the pathogenesis of myofibroma may be associated with mutations in the *PDGFRB* and *NOTCH3* genes. The literature indicates that subsets of infantile myofibromatosis (a subtype of myopericitoma-like lesions) occur in families according to autosomal dominant inheritance, although some reports suggest autosomal recessive inheritance [1].

The diagnosis of myofibroma is possible based on histopathological examination. Morphological characteristics in imaging studies, patient's age, size of lesions and their number are helpful in making the diagnosis [6]. Macroscopically, myofibromas are painless, rapidly growing soft tissue tumors that can cause bone erosion [2, 3]. Histologically, myofibroma consists of myofibroblasts and undifferentiated cells distributed around vessels, resembling hemangiopericytoma [3].

Imaging diagnostics is an essential element of the procedure and, in addition to the possibility of making a preliminary diagnosis, allows for precise determination of the location and dimensions of the lesion and the extent of invasion of adjacent anatomical structures, which is crucial for further treatment. The literature contains features that can be observed in the case of myofibroma using imaging diagnostics methods.

According to the literature, the echogenicity of myofibroma on ultrasound is variable, and the lesions themselves may contain fluid spaces corresponding to areas of necrosis or hemorrhage.

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In addition, calcifications and a varying degree of vascularization in the Doppler option may occur within the lesions. Another key imaging method that allows the identification of a proliferative lesion as myofibroma is magnetic resonance imaging (MRI). In MRI studies, myofibroma may demonstrate an isointense signal in relation to muscles in T1-weighted images. It usually also demonstrates a hyperintense signal in T2-weighted images with areas of necrosis or hemorrhage. Enhancement after administration of a gadolinium-based contrast agent may be diffuse or inhomogeneous and may demonstrate a target sign due to necrotic areas in the central part [3, 6, 7, 8].

Differential diagnosis in imaging studies, in addition to the morphology of the lesion, also requires taking into account the patient's age and the presence of disease foci in other anatomical locations. According to the literature, infantile myofibromatosis-type lesions require differentiation from a spectrum of lesions, which mainly include hemangioma, congenital infantile fibrosarcoma, neurofibroma, soft-tissue sarcoma, metastatic neuroblastoma, and histiocytosis [6].

In the presented case, the authors discuss the context of imaging diagnostics of a myofibroma lesion that was confirmed histopathologically and consider the possible relationship between the appearance of the lesion and the previous trauma.

CASE REPORT

The presented case concerns a 3.5-year-old boy whose parents observed a rapidly growing tumor in the frontal region during a week and a half before the child was admitted to the hospital for further diagnostics. The interview revealed that the boy had suffered a head injury 2 months earlier, in the form of being hit by a swing. Since the injury, the frontal region has been under periodic surgical observation due to a suspected post-traumatic hematoma. During the admission to the hospital, a physical examination revealed a tumor in the frontal region measuring approx. 6 x 6 cm, which involved the left orbit and pressed against the eyeball. In the clinical assessment, the tumor was painless, but the boy was unable to open his left eye independently.

The initial ultrasonud examination of the soft tissues of the frontal region (Fig. 1) revealed a poorly demarcated, non-movable, non-compressible mass with approximate dimensions of 3 x 5 x 5 cm. The lesion was filled with heterogeneous, hypoechoic content (solid or fluid); hyperechoic bands were visible within the lesion near the squamous part of frontal bone, which could correspond to calcifications or bone destruction. The Color Doppler examination did not clearly demonstrate vascularization within the lesion. Finally, the assessment of the lesion in the ultrasound examination showed that the morphology of the lesion was non-characteristic, while in the differential diagnosis, apart from a hematoma in the process of evolution (due to the post-traumatic background in the interview), appeared a proposition also to consider histiocytosis or another proliferative background, with a suggestion of extending the diagnostics.

The patient also underwent a MRI scan of the head (Fig. 2), which revealed a tumor in the left frontal region measuring

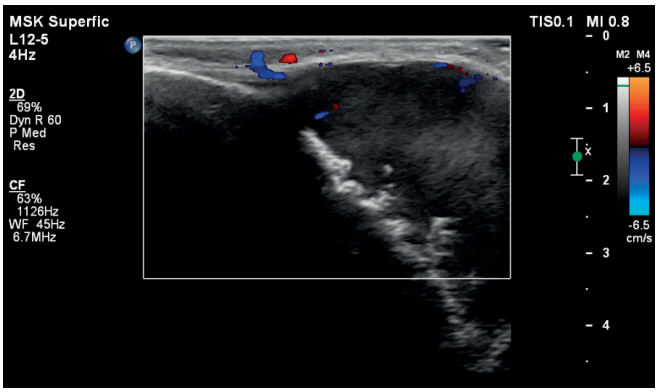

FIGURE 1. Ultrasonographic image of the lesion showing the changed cortical layer of the skull and the lack of significant vascularization of the lesion in the Color Doppler option

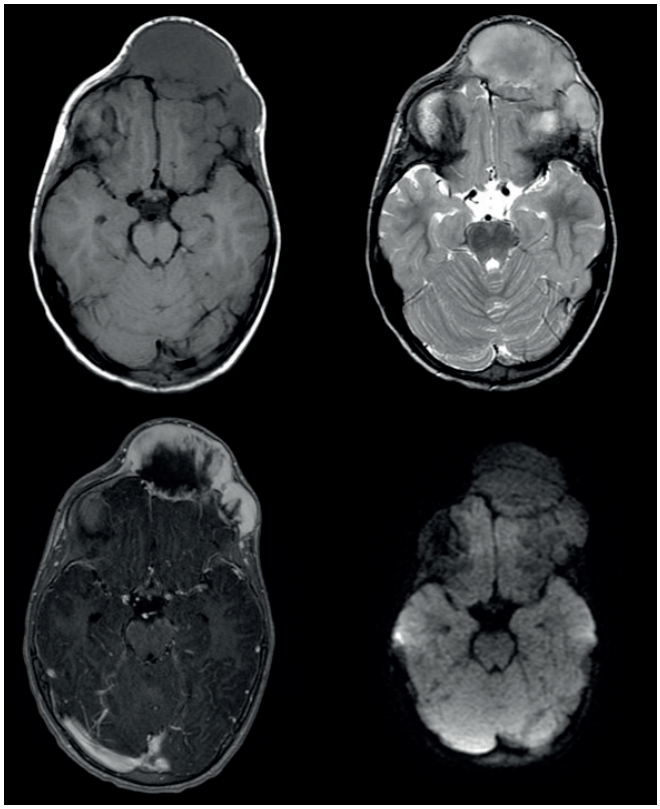

FIGURE 2. Image of the lesion in the magnetic resonance imaging (MRI) examination in the following sequences: T1 – weighted images (top left), T2 – weighted images (top right), T1 – weighted images after contrast agent administration (bottom left) and diffusion-weighted imaging – DWI (bottom right)

56 x 39 x 52 mm, with intermediate signal intensity in T1- and T2-weighted images, which was contrast enhanced mainly in the peripheral parts of the lesion and did not show signs of restricted diffusion on diffusion-weighted imaging (DWI) sequence. The larger component of the tumor was located extracranially and was limited by the tendon cap. The tumor caused destruction of the external and internal lamina of the frontal bone and penetrated intracranially, where it seemed to be limited by the dura mater of the brain. The lesion modeled the base of the left frontal lobe and filled the upper part of the left orbit downward, displacing the eyeball downward. Susceptibility-weighted imaging (SWI) images revealed small signalless foci in the tumor

mass, which corresponded to calcifications or post-hemorrhagic hemosiderin deposits. In addition, another intracranially penetrated lesion, measuring 26 x 21 x 26 mm, was visible lateral to the largest lesion. Also several smaller foci of similar signal intensity and contrast enhancement, of up to 12 mm, were also visualized in the frontal bone on both sides. The assessment of lesions in the MR examination indicated that the spectrum of differentiation of the described changes is quite wide, and differential diagnosis should include diagnoses such as histiocytosis, sarcoma and metastatic changes.

During the further course of hospitalization, a sample of the tumor was taken and the material underwent histopathological examination. Histopathological evaluation showed that the lesion was composed of elongated, spindle-shaped cells, without clear cytological features of malignancy. The microscopic image indicated a benign lesion of mesenchymal origin (possibly cranial fascitis), with the suggestion that the final histopathological diagnosis would be established after removal of the entire lesion for histopathological examination. During hospitalization, the tumor was observed to grow in size. The boy was qualified for surgery to resection the lesion. Frontal craniotomy and left orbitotomy were performed. The tumor was removed from the anterior cranial fossa, left orbit and ethmoid. Fragments of bone infiltrated by the tumor were also removed and cranioplasty was performed. Histopathological analysis of the obtained material revealed a spindle cell tumor with the presence of perivascular systems (hemangiopericytoma-like). The tumor infiltrated fragments of bone tissue. The final histopathological diagnosis of the removed lesion was myofibroma/myofibromatosis. Immunohistochemical tests showed the following results: EMA (–), CD34 (–), Vimentin (+), S-100 (+ in few cells), SMA (–), GFAP (–), Olig2 (–), p53 1–3%, Ki67 up to 25%.

A follow-up MRI of the head, performed 2 weeks after the surgery, revealed several foci of strongly enhancing, pathological tissue infiltrating part of the left frontal lobe, the medial part of the left eyeball, the oculomotor muscles of the left eye, the left ethmoid sinus and the cavernous sinus. In addition, small foci of similar morphology in the frontal bone were suspected of infiltrating the meninges and the frontal lobe. A computed tomography (CT) scan of the chest and abdomen did not reveal foci suspicious of myofibromatosis in these anatomical areas. Apart from surgical treatment, during the entire treatment at the local clinic, the patient received 5 cycles of cytostatic treatment (vincristine) with good tolerance. Then the patient and his family changed their place of residence, hence further treatment and monitoring of the patient was taken over by another medical facility.

DISCUSSION

In the literature, one can find information about various methods of systemic treatment of multifocal myofibromatosis, also with varying results. Vincristine was used in high doses for inoperable tumors or for tumor recurrence after surgery. The risk of recurrence with vincristine and dactinomycin therapy was described as low, with recurrence reported in 1 in 9 cases [9]. The commonly recommended treatment for myofibroma is surgical resection. It is also important to regularly monitor patients for possible recurrence. In a study published in 2017, tumor progression was noted in 1 of 42 patients during a follow-up of 50.5 months [10, 11].

Although infants with multicentric or generalised myofibromatosis involving the viscera and brain have a poorer prognosis, the overall prognosis for solitary myomas is good. Most myofibromas do not recur, even if marginally or incompletely resected, and aggressive growth with distant metastases of myomas in a malignant form is considered extremely rare [1].

The association of a previous injury with the occurrence of myofibroma-type lesions in soft tissues and bones appears frequently in the medical literature and could be a valuable subject of extended research, which, however, is certainly hampered by the rare occurrence of these lesions. Cases of orbital myofibroma in children can also be found in current medical literature, but there is no traumatic context in the cases that the authors managed to access [5].

The literature describes a case of a 29-year-old woman whose history revealed regular biting of the cheek, where a lump was diagnosed. Microscopic analysis of the lesion suggested the occurrence of a fibroma [12].

It is also worth mentioning the case described by Akdağ, concerning a fibroma diagnosed in a 26-month-old girl. In this case, according to the interview with the family, 45 days earlier the child had suffered a head injury, after which she was under otolaryngological and surgical observation. The child was initially diagnosed with a hematoma and an incision was made to drain the blood. Due to persistent swelling of the cheek and deformity of the nose, the girl's family and the child reported to the outpatient clinic. The child underwent a rhinoscopic examination and CT, which showed the presence of a mass in the right maxillary sinus, which spread towards the infraorbital region. The mass was subjected to surgical extraction, and histopathological analysis showed the presence of a fibroma. In discussing the possible origin of the lesion, the author suggests speculating on the possible role of trauma as a critical trigger that could have allowed uncontrolled growth of a nonaggressive tumor that may have been present before the traumatic event. However, he also notes that this topic requires further investigation using meta-analysis or clinical trials with a large number of patients [13].

Dhupar et al. in their publication described a case of intraosseus myofibroma of the jaw of 9-year-old girl, which occurred after the extraction of the left second primary molar of the mandible 4 months earlier due to severe caries. In their publication, the authors, in their considerations of the hypothesis of the origin of the lesion, emphasize the role of myofibroblasts in supporting wound healing and mention the possible proliferation of myofibroblasts in the area affected by the injury [14].

Example of posttraumatic occurrence of intraosseous myofibroma is presented by Santos et al. in their publication, where they describe the case of a 15-year-old girl who experienced a physical assault with facial injuries. A month after this event, the patient experienced severe pain and difficulty in chewing. Cone beam computed tomography showed features suggestive of a pathological fracture associated with a lesion in the mandible. After surgical treatment, no signs of recurrence of the lesion were observed in the patient [15].

CONCLUSIONS

In the context of the above-mentioned reports, the authors of this publication are inclined to assume that the injury that occurred in the boy in the described case could have been related to the later appearance of a myofibroma in the frontal and orbital regions.

Moreover, this case suggests emphasizing the value of control ultrasound examinations, which are the first and most easily accessible to assess the evolution of post-traumatic changes in paediatric patients. Ultrasound control of the post-traumatic area, in addition to observing the evolution of a possible hematoma, should also encourage oncological vigilance for bone destruction. The lack of regression of the dimensions of the lesion (and especially their progression) should be an indication for urgent extension of diagnostics.

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