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Commentary on "Primary orbital monophasic synovial sarcoma with calcification: A case report"

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Abstract
The present letter to the editor is related to the study titled “Primary orbital monophasic synovial sarcoma with calcification: A case report”. Orbital synovial sarcoma is one of the rare intraorbital masses seen in adult and pediatric populations. Some case reports in the literature revealed synovial sarcoma may contain calcifications. Therefore, it is important to make differential diagnosis among calcified orbital masses in childhood.

TO THE EDITOR
We read the article “Primary orbital monophasic synovial sarcoma with calcification: A case report”[1] with great interest and appreciated the authors for this comprehensive case report. We also thought that it might be favorable to contribute additional information about differential diagnosis and shortly immunohistochemical features to the discussion. For this purpose, we focused on the differentiation among the pediatric intraorbital calcific masses.

In the literature, intraocular [2] and extraocular [3-5] synovial sarcoma cases have been reported. Retinoblastoma is one of the most common intraocular tumors with calcification in children under 5-year-old. The presence of calcification is an essential feature [6]. It shows hypointense on T2 gravimetric imaging (WI), and slightly hyperintense on TIWI on magnetic resonance imaging (MRI) compared with the vitreous. Besides, heterogeneous enhancement can be seen on post-enhanced imaging.
This case report reported introocular synovial sarcoma in a 48-year-old female patient \[2\] and retinoblastoma was not included in the differential diagnosis due to a possible age factor.

Rhabdomyosarcoma is one of the relatively more common masses in children. On computed tomography (CT), it is usually seen as an extraconal irregular ovoid, well-circumscribed mass. If there is destruction on adjacent bone, concurrent calcification can be seen. As its size increases, it becomes more heterogeneous and its borders are unclear. The eyelid thickening is a typical finding even without an extension. On MRI, it is hypointense on T1WI and hyperintense on T2WI \[7\].

Synovial sarcomas should also be differentiated from metastases. The most common pediatric orbital metastases are neuroblastoma. The presence of a primary tumor in the retroperitoneum or posterior mediastinum would facilitate the diagnosis \[7\]. Hyperdense appearance of neuroblastoma metastases in CT series is also helpful in differential diagnosis \[7\]. Ewing sarcoma metastasis can also be considered in children. Immunohistochemical features are helpful in differentiating Ewing sarcoma from the synovial sarcoma. EMA and CK7 are helpful in diagnosing synovial sarcoma, while CD99/Fli-1 is helpful in Ewing's sarcoma \[8\]. In addition, calcification can be seen as a result of dystrophic calcification in metastatic tumors, unlike the others \[3\].

Dermoid cyst is one of the most common orbital masses in children. Since it may contain calcification, it should be included in the differential diagnosis of synovial sarcoma. Bone changes may be the cause. The cystic component, fluid levels and the presence of fat attenuation (associated with T1 high signal on MRI) are helpful in the differential diagnosis \[7\]. In addition, diffusion restriction on diffusion weight imaging (DWI), non-enhancement in post-contrast images, and smooth contours can aid in differential diagnosis \[6\].

Infantile hemangioma is the most common tumor in infancy and although calcification is rarely present, it should be considered in the differential diagnosis. It is usually located extraconally and makes some changes adjacent bone-like expanding or scalloping but invasion occurs extremely rare. It enhances homogenously after contrast
administration. On T1WI, the well-defined marginated mass is often iso- to hyperintense compared to muscle, and moderately hyperintense on T2WI with flow voids within the tumor. The presence of a flow void is an important feature to differentiate from the other masses [7].

Meningiomas account for 2% of primary orbital tumors and they are caused by the periosteum of the orbital wall. It may show coarse diffuse calcifications and sclerosis in the optic foramen that are helpful in the diagnosis. Although not specific, central radiolucent line may be seen [3, 6].

Peripheral nerve sheath tumor (PNST) is one of the calcified intraorbital tumors. Histopathologically, it can express S100, EMA, CK7, CK19 TLE 1 and SOX10 as synovial sarcoma. On the other hand, while PNST expresses CD34, it is rarely seen in synovial sarcoma [3, 9].

Finally, we could contribute to the current study about immunohistochemical features of synovial sarcomas. They nearly all express EMA (+), cytokeratin (especially CK 7) (+) and 30% of them express focal S100 (+). CD99 (+) is also expressed in 60-70%, and LTE1 (+) is >90%. In contrast, CD34 is rarely/ seldom expressed. The current study presents EMA, CK 7, S-100 are negative and CD34 is positive in immunohistochemical study, unlike the previous studies [3, 5, 9].