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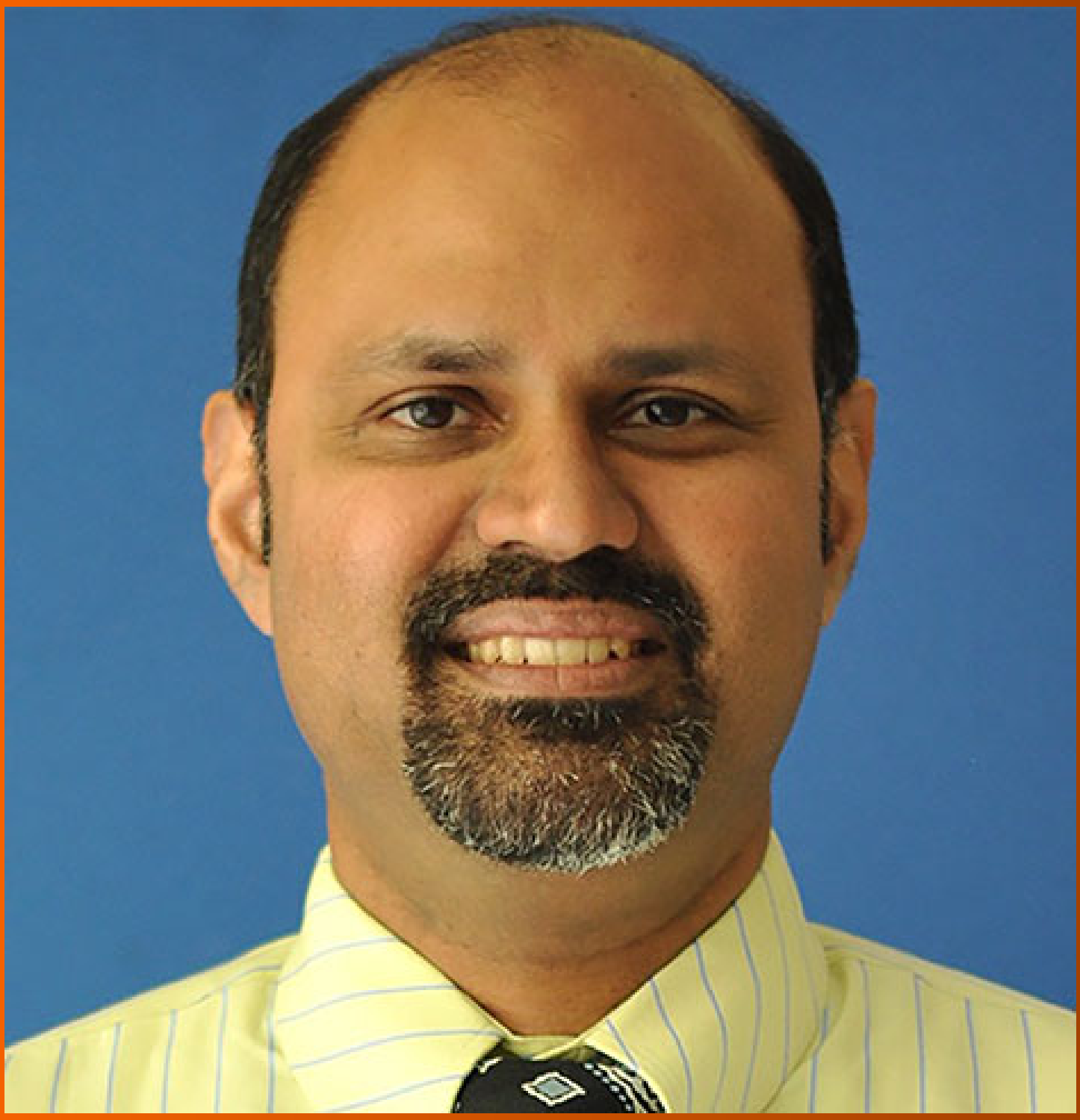

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LETTER TO THE EDITOR

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

Oguzhan Tokur, Sonay Aydın, Erdal Karavas

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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 that synovial sarcoma may contain calcifications. Therefore, it is important to make differential diagnosis among calcified orbital masses in childhood.

Key Words: Orbital tumor; Synovial sarcoma; Calcification; Children; Histopathology; Radiology

Core Tip: This letter to editor serves to contribute additional information regarding differential diagnosis and immunohistochemical features to the article. We hope that by using radiographic and immunohistochemical features, we can assist in differentiating calcified orbital masses in the pediatric population.

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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 is hypointense on T2 gravimetric imaging (WI), and slightly hyperintense on T1WI on magnetic resonance imaging (MRI) compared with the vitreous humor. 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 the 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 adjacent bone destruction, 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 on 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 high T1 signal on MRI) are helpful in the differential diagnosis[7]. In addition, diffusion restriction on diffusion weight imaging, 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 to adjacent bone like expanding or scalloping, but invasion occurs extremely rare. It is enhanced homogenously after contrast administration. On T1WI, the well-defined marginated mass is often isointense 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 (+) and cytokeratin (especially CK 7) (+), and 30% of them express focal S100 (+). CD99 (+) is also expressed in 60%-70%, and LTE1 (+) occurs in > 90%. In contrast, CD34 is rarely/seldom expressed. The current study presented that EMA, CK 7, and S-100 were negative and CD34 was positive in immunohistochemical study, unlike the previous studies[3,5,9].

FOOTNOTES

Author contributions: Tokur O and Aydin S contributed equally to this work; Tokur O, Aydin S, and Karavas E designed the letter; Tokur O and Aydin S performed the research; Tokur O wrote the manuscript; all authors have read and approved the final manuscript.

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