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Case Report

Composite Hemangioendothelioma Settled in the Paraspinal Region: A Rare Case Report

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ABSTRACT

Composite hemangioendothelioma (CHE) is a very rare, low-grade malignant vascular tumor. It is most commonly seen as one or more nodules on both sides of the fingers and toes. This tumor rarely settles in the paraspinal muscles. The age of onset is usually between 21 and 72 years, with an average age of 39.5-41 years. Treatment is with gross total removal of the lesion. We present a case of a 54-year-old male patient with a 2-year history of low back pain. Lumbar spine magnetic resonance imaging revealed CHE in the right paravertebral muscle in the posterior vicinity of the transverse process between the right paraspinal muscle planes. The entire mass was removed grossly. The definitive diagnosis of CHE was made histopathologically. This is a rare case of CHE that had settled in the paraspinal region.

KEYWORDS: Composite hemangioendothelioma, Paraspinal region, Spine

ABBREVIATIONS: CHE: Composite hemangioendothelioma; **HE:** Hemangioendothelioma

INTRODUCTION

Composite hemangioendothelioma (CHE) is a recently described, very rare vascular tumor with borderline low-grade malignancy (6,12,14). It is most commonly seen as one or more nodules on both sides of the fingers and toes (6). The age of onset is usually between 21 and 72 years (6), with an average age of 39.5–41 years (6,8). Other types of hemangioendotheliomas (HEs) are similar to CHE, but are histologically very different (6). CHE shows strong morphological similarities with benign vascular tumors, including spindle-cell HE such as epithelioid HE and retiform HE (6). CHE cases have been reported by Nayler et al.(12) and two other linked studies (4,13).

CASE REPORT

Our patient was a 54-year-old man with a 2-year history of low back pain for which he had undergone various medical treatments and physical therapy. The patient was referred to our outpatient clinic for lumbar magnetic resonance imaging (MRI). The results of neurological examination were normal. MRI revealed a heterogeneous lesion of 26 mm in diameter that was hyperintense on T2-weighted (T2-W) images (Figure 1A) and minimal hyperintense relative to the muscle on T1-weighted (T1-W) images (Figure 1B). A massive heterogeneous and contrast-enhanced tumoral lesion was observed. Lumbar spinal computed tomography (CT) was performed to observe the spinal bone structures. CT revealed a heterogeneous and contrast-enhanced nodular lesion 3 cm in diameter along with surrounding fat tissue in the right paraspinal muscle at the L4

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level (Figure 1C). The patient underwent surgery, and a lesion 3×2.5×2 cm in size was removed from the right paraspinal muscle in the right lumbar region.

Pathological examination of the lesion showed that it consisted of various components. There were large or small ectatic thin-walled capillary structures in its periphery, especially in non-lesion areas. There were thick-walled vascular structures whose luminal endothelium had an epithelioid appearance and that bulged toward the lumen (Figure 2A). The lesion was generally composed of a spindle-cell proliferation showing minimal or moderate atypia. Spindle cells formed vascular slit-like spaces with a large number of erythrocytes. There were signs of previous bleeding in some places, which were

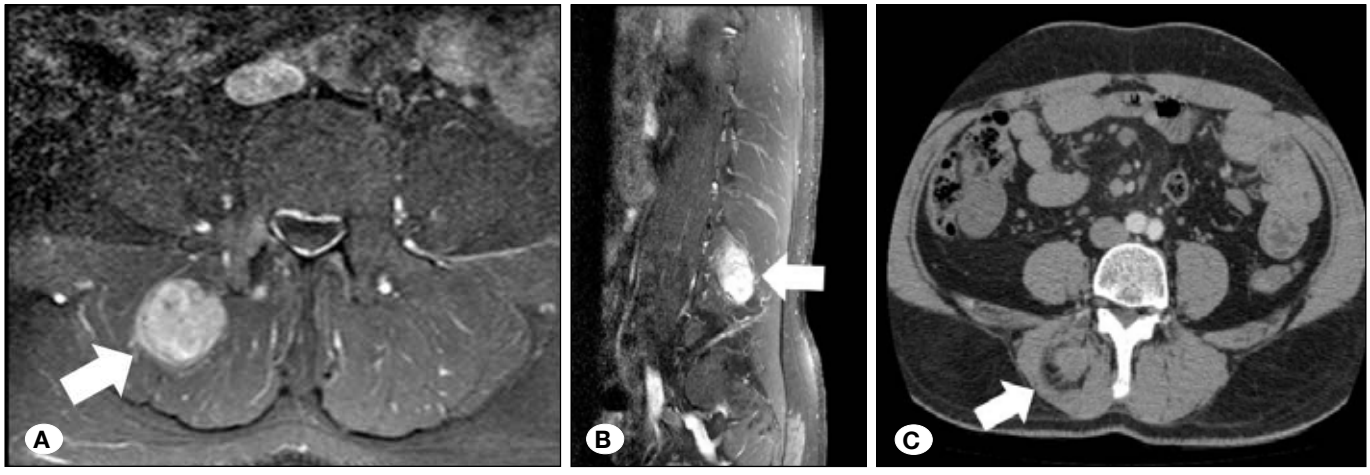

Figure 1: A) Heterogeneous hyperintense lesion in T2-W axial MRI, 26 mm in diameter, neighboring transverse process posterior (white arrow) between right paraspinal muscle plans. **B)** In T1-W sagittal MRI scan, minimal hyperintense lesion is observed when compared with the muscle (white arrow), and there is intense heterogeneous contrast enhancement. **C)** In axial lumbar CT, at L4 level, in the right paraspinal muscle, there is a nodular mass lesion (white arrow) that enhanced heterogeneously with contrast with a diameter of 3 cm, and there is also fat tissue surrounding it.

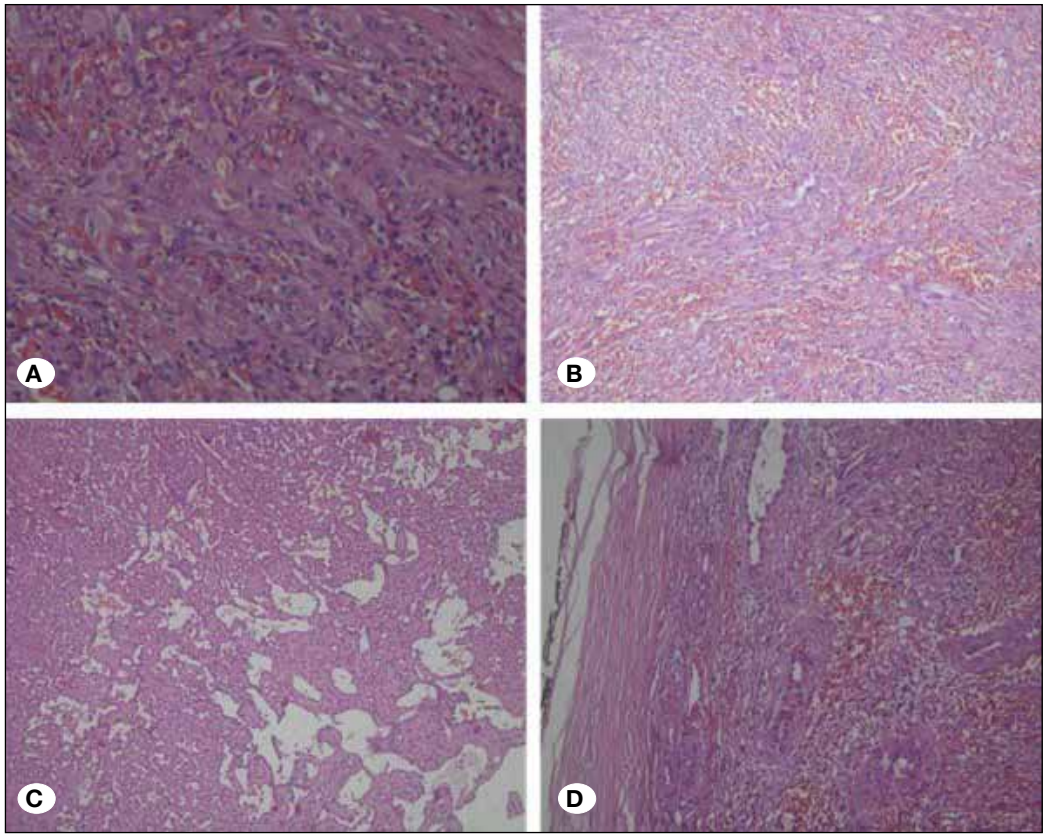

Figure 2: A) Composite hemangioendothelioma. This area is consistent with angiosarcoma (H&E, x400). **B)** Composite hemangioendothelioma. This area is consistent histologically with Kaposi sarcoma (H&E, x200). **C)** Composite hemangioendothelioma. This area is consistent with retiform hemangioma (H&E, x100). **D)** Composite hemangioendothelioma. Epithelioid hemangioma-like area (H&E, x200).

interpreted as Kaposi sarcoma-like areas (Figure 2B). Another part of the tumor had vesicular hypochromic epithelioid cells that had a very large and pale eosinophilic cytoplasm, occasionally contained cytoplasmic vacuoles, and had large nuclei (Figure 2D). These cells formed cell cords showing subsequent sequences in loose stroma and abortive vascular structures associated with these cords. These areas were consistent with epithelioid HE. In some areas, epithelioid cells formed retiform and papillary structures with fibrotic cores as seen in retiform HE (Figure 2C). Lipoblast-like vacuolar cells were present in a few areas, and some areas were quite hypocellular. The uniform appearance of spindle cells in this area was similar to spindle-cell hemangioma.

Immunohistochemical analysis revealed that the tumoral tissue was widely and strongly stained with CD31 and showed focal membranous staining with CD34. While there was moderate staining with thrombomodulin, only a small number of thin-walled ectatic vascular structures were stained with podoplanin. HHV8, desmin, and EMA were negative. Very few cells were stained with CK7. There was intense staining in actin filaments and around the vascular structures in most of the stroma. Although the Ki67 index was 18%–20%, especially in areas where the tumor was cellular, the index was generally considered to be around 10%.

DISCUSSION

HE was used to identify vascular lesions showing biological and morphologic differences. According to the World Health Organization (WHO) classification, mesenchymal tumors and HEs have been classified as moderately differentiated borderline vascular tumors. CHE is a low-grade tumor that rarely results in metastasis and death (6), although it often recurs. A distant metastasis was not an expected finding and none were detected at the 1-year follow-up of our patient. HEs include spindle-cell HE, CHE, papillary HE, Kaposi-form HE, retiform HE, epithelioid HE, and polymorphic HE (11); CHE is very rare (4,12). The cases reported in the literature are single or multinodular poorly circumscribed lesions on the hands and/or feet of adults (6). CHE is the most recently described HE type and was first described as a low-grade tumor by Nayler et al. in 2000. Eight CHE cases have since been reported (12). In 7 cases, CHE was located in the dermis and subcutaneous tissue; in one case, oral mucosal involvement was found. Fukunaga et al.(9) reported that CHE is composed of a complex admixture of benign, intermediate, and malignant vascular components. They reported that epithelioid and retiform HEs were predominant, including a mixed histopathology variant in two of the tumors. Aydıngoz et al.(3) reported CHE in some regions of lesion recurrence 3 months after excision and electrocauterization of a pyogenic granuloma in the thigh. In our patient, CHE was located in the paraspinal region, which is very rare. Metastatic carcinomas (e.g., hemangioma, multiple myeloma, telangiectatic osteosarcoma, Ewing sarcoma, and renal cell carcinoma) and infectious diseases should be considered in the histopathological differential diagnosis (16).

Cases of spontaneous vascular malformation and hemangioma development have rarely been reported in the literature (7,10), Rossi and Fletcher (15) reported four cases. Recent experimental studies on the pathogenesis of angiosarcoma demonstrated a malignant transformation in hemangiomas (1). P53 and vascular endothelial growth factor (VEGF) have been shown to play roles in the pathogenesis of angiosarcoma (19), and a hereditary relationship with soft-tissue angiosarcoma has been reported (5). Although the development of a melanoma and metastatic tumor from an intravascular lymphoma and hemangioma has been reported (17,18), vascular endothelial cell markers (CD31, CD34, and FVIII) provide the distinction by positive immunostaining of vessels (6). In our patient, immunohistochemical analysis revealed that the tumoral tissue was widely and strongly stained with CD31 and showed focal membranous staining with CD34. While there was moderate staining with thrombomodulin, only a small number of thin-walled ectatic vascular structures were stained with podoplanin. Because HE is a vascular tumor, the possibility of perioperative blood loss is high (2). In our patient, one unit of erythrocyte suspension was administered because 700 mL of blood was lost peroperatively.

CONCLUSION

CHE is a rare low-grade vascular tumor that is usually seen in the skin and subcutaneous tissue. It rarely settles in the spine or extremities. CHE in the paraspinal region has not been reported in the literature and should be considered in the differential diagnosis of lesions located in this region.

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