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CASE REPORT

Primary intracranial hemangiopericytoma with mesenteric and retroperitoneal spread

Yesilkaya Y*, [1], Akinci D[1], Topcuoglu M[1], Tanas O[2], Altundag K[3]

1 Department of Radiology, Hacettepe University School of Medicine, Ankara, Turkey
2 Department of Pathology, Hacettepe University School of Medicine, Ankara, Turkey
3 Department of Medical Oncology, Hacettepe University School of Medicine, Ankara, Turkey

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ABSTRACT

Hemangiopericytoma is an uncommon mesenchymal tumour that originates from Zimmermann's pericytes. It accounts for about 0.4 % of all tumours that occur within the primary central nervous system (CNS). It is characterized clinically by its delayed local recurrence and extracranial metastases. To the best of our knowledge, mesenteric and retroperitoneal metastases from meningeal hemangiopericytomas have not been reported before. Herein we present the case of a 60-year-old man who exhibits mesenteric and retroperitoneal metastases from primary intracranial hemangiopericytoma after 11 years of follow up.

Keywords: Intracranial hemangiopericytoma, Metastasis, Mesenteric, Retroperitoneum

INTRODUCTION

Hemangiopericytoma is a rare vascular tumour that was first described by Stout and Murray in 1942. This tumour originates from the pericytes of Zimmerman. It most frequently occurs in the fourth and fifth decades and there is no gender predilection [1–8]. Owing to the ubiquity of pericytes in mesencyhmal tissues, hemangiopericytomas may occur anywhere in the body although they are most commonly found in the retroperitoneum, pelvic fossa, extremities, head and neck [3]. The intracranial localization is not frequent [4, 5]. It accounts for approximately 0.4 % of all primary central nervous system (CNS) tumours [5, 6]. It is commonly accepted that hemangiopericytoma may present with delayed local and distant recurrences [3, 6]. The lungs are the most common site for metastasis followed by bones and the liver [3, 4].

* Corresponding author. Address: Department of Radiology, Hacettepe University Medical School, Sihhiye Ankara 06100, Turkey. Tel.: +90-312-305 11 88; Fax: +90-312-311 21 45; E-mail: dryakup23@hotmail.com (Yakup Yesilkaya)

CASE REPORT

A 60-year-old man was admitted to our hospital in 2000 complaining that he had suffered from two weeks of tinnitus. Initial physical examinations indicated that he was healthy and no obvious cause of the tinnitus could be identified. Brain magnetic resonance imaging revealed a large heterogenous right frontotemporoparietal mass that measured 8×7 cm in diameter. The tumour was surgically removed. A histological examination confirmed that the tumour was hemangiopericytoma. The patient had undergone

primary surgery for a right frontotemporoparietal region mass in 2000, which had been followed by adjuvant radiotherapy. Nine years after surgery, three separate intraabdominal tumours were detected by ultrasonography and computed tomography. These tumours were identified as hypoechoic masses with a mean tumour size of 20 mm (Figure 1). They were located next to the right kidney (Figure 2a), next to left kidney (Figure 2b) and in the right lower abdominal quadrant (Figure 2c). Contrast-enhanced computed tomography depicted them as well-defined masses with homogeneous contrast enhancement without calcifications. A CT scan of the chest showed the presence of multiple metastatic nodules in the lung (Figure 3). The second biopsy material of the patient was obtained from intra-abdominal nodule using a fine needle procedure. A histopathological examination revealed a hypercellular neoplasm with hemangiopericytic vascular pattern (Figure 4), which was highlighted with reticulin stain. Neoplastic cells had oval to spindle pleomorphic nuclei with prominent nucleoli and syncytial eosinophilic cytoplasm. The mitotic rate was low. Immunohistochemical studies showed that neoplastic cells were immunoreactive for CD34 and vimentin, and negative for EMA (epithelial membrane antigen). The microscopical examination of this biopsy demonstrated similar morphological and immunohistochemical features of the first biopsy. An ultrasound-guided biopsy confirmed the diagnosis of late metastases from the primary intracranial hemangiopericytoma. The patient received 3 cycles of ifosfamide and adriamycine combination chemotherapy. At the end of 3 cycles, the disease was stable and the patient underwent a further 3 cycles of treatment. The patient survived the cancer and follow-up examinations indicate that he has not experienced any further problems.

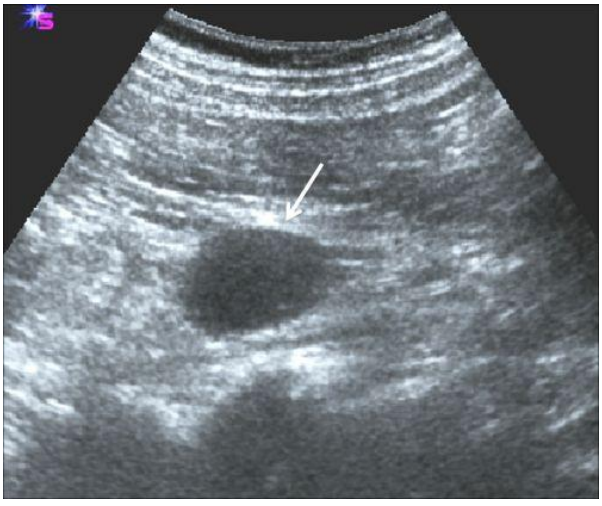
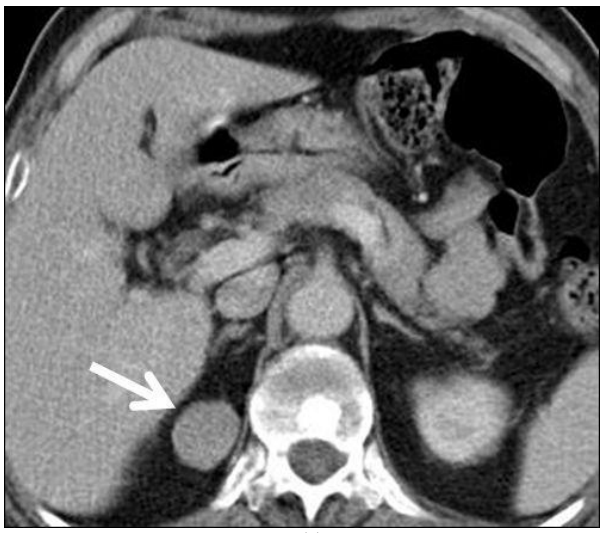
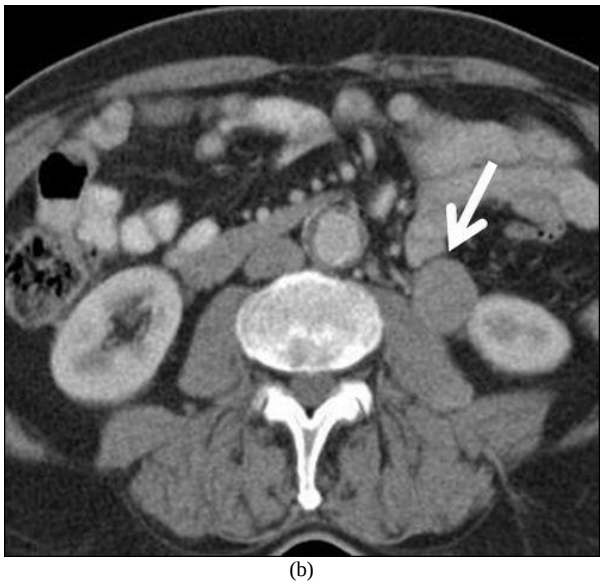


Figure 1 A US examination revealed a solid hypoechoic mass in the mesenteric tissue.



(a)



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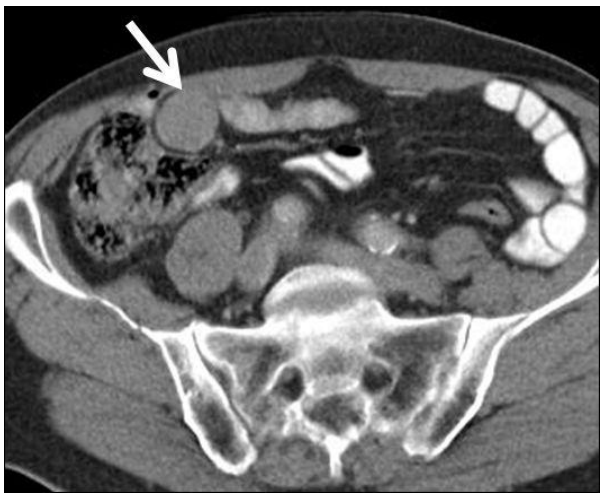

(c)

Figure 2 Contrast-enhanced computed tomography depicting mesenteric and retroperitoneal masses.

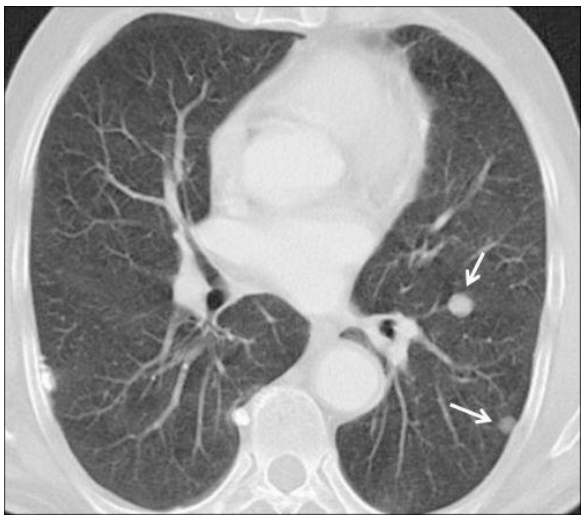

Figure 3 Computed tomography showing the multiple metastatic nodules in the lung.

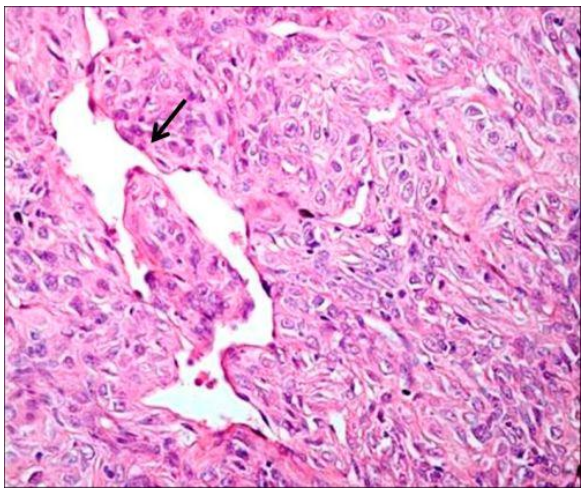

Figure 4 Histopathological examination revealing a hypercellular neoplasm with hemangiopericytic vascular pattern.

DISCUSSION

Malignant hemangiopericytomas represent a small group (2–4%) of meningeal tumours; unlike meningiomas, they may show local recurrence and extraneural metastases [5]. Meningeal hemangiopericytomas are fast growing tumours and they are rich in vascularity. Hemangiopericytoma has a stronger tendency to recur and metastasize than meningioma. The recurrence and metastasis rates of intracranial hemangiopericytoma were reported to be 80 percent and 23 percent respectively [2]. The prognosis of hemangiopericytoma is generally favourable, although recurrent tumours or metastatic tumours or both develop in some patients after surgical treatment. Enzinger at al. reported that 17% of 93 patients suffered from metastases and 14% died of the disease. They also reported a 10-year survival rate of 70% [3]. Interestingly, the interval to disease recurrence is longer than that reported for other sarcomas and metastases can develop following an extended disease-free interval, possibly contributing to the favourable prognosis [3]. Conventional morphologic criteria for distinguishing a benign hemangiopericytoma from a malignant tumour are helpful for predicting the prognosis. The combination of a tumour larger than 5 cm in diameter, prominent mitotic activity, (more than 4 mitoses per 10 high-power fields) and tumour cell necrosis is reported to be strongly indicative of malignant hemangiopericytoma [3]. Several previous reports have suggested that prognosis is related to the size of the tumour [3]. Extracranial metastases occur in 23 percent of cases and appear at mean of 8 years after the initial therapy [4]. The liver, lung and bone are the most common sites of metastases [4]. Anecdotal pancreas, spleen and heart localizations have been reported, but this is the first case in the literature where mesenchymal and peritoneal metastases have been documented. The radiologic appearance of HPC, primary or metastatic, is non-specific. The imaging findings of hemangiopericytoma are similar, regardless of whether the tumour is intra- or extracranial. In a report on multiple hemangiopericytomas, the lesions were seen as hypoechoic nodules on sonograms. Alpern *et al*. described a small series of primary abdominal HPCs and described CT findings of a lobulated mass with enhancing solid components, cystic areas and speckled calcification as ‘suggestive of, but not specific for, hemangiopericytoma. Calcifications and cystic areas were less commonly found in other series’ [5, 8, 9]. The identification of a highly vascular mass appears common among all reports. HPCs are frequently described as well developed vascular networks with a dense capillary blush on angiography and intense enhancement on CT [8, 9]. However, the patient in this case did not exhibit an intense contrast enhancement and this may be of significance. Surgical excision with curative intent is the standard strategy for locally recurrent masses, distant metastatic lesions and primary tumours, and is associated with an overall favorable outcome [3]. Since most recurrences developed at distant sites, beyond the scope of surgical resection, systemic adjuvant chemotherapy may be an option for malignant hemangiopericytoma [7]. Wide excision is the treatment of choice for primary malign hemangiopericytomas. However, despite the accomplishment of radical surgery, local recurrences occur in as many as half of the cases [5]. Surgical excision and postoperative radiotherapy at a dose of 50 Gy or more is recommended as the initial treatment, as adjuvant radiotherapy results in significantly better local control than surgery alone [4]. Past results indicate that local treatment is strongly related to the final prognosis. However, even though the best local treatment has been performed, the prognosis remains dismal due to the unpredictable malignancy of hemangiopericytomas [1]. The management of local recurrences can be surgical, radiosurgical or radiotherapeutic.

CONCLUSION

In summary, careful life-long follow-up is required for patients with hemangiopericytoma, as recurrence and

metastases can develop after an extended disease-free interval.

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