Brain Metastasis Of Penile Angiosarcoma

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Angiosarcoma is a rare malignancy originating from vascular endothelial cells. Brain metastasis of aniosarcomas are uncommon up to the literature. Penile angiosarcomas are also seldom among all anjiosarcomas. A case with penile angiosarcoma with confirmed brain metastasis is aimed to be reported and contribute to the literature for similar cases.

INTRODUCTION

Angiosarcoma is a very rare, agressive malignancy originating from vascular endothelial cells. It can be primarily seen on the head, face, liver, skin and other soft tissues.(1) Penile located angiosarcoma is very rarely seen.(2,3) Its treatment is generally surgical with adjuvant chemotherapy or radiotherapy.(4) Brain metastasis is an unusual condition in angiosarcomas, therefore very few cases have been described.(1) Our aim is to present a rare case of penile angiosarcoma with brain metastasis. The case contributes to the literature on the diagnosis, treatment and treatment process.

CASE REPORT

A 35-year-old male patient was admitted to the urology clinic with infiltrative mass on the glans penis. On ini-

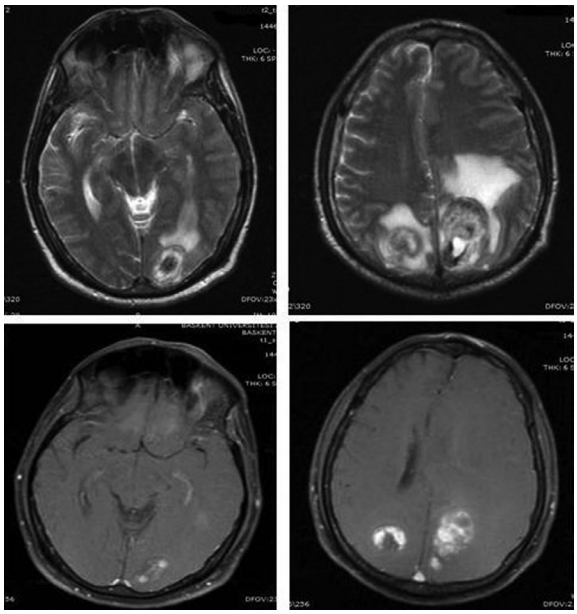

Figure 1. T2 and T1 weighed enhanced axial MR images showing metastatic mass lesions. largest one is localized on the left parietal cortical-subcortical area and all masses are heterogeneously enhanced with hemorrhagic content and concominant vasogenic edema.

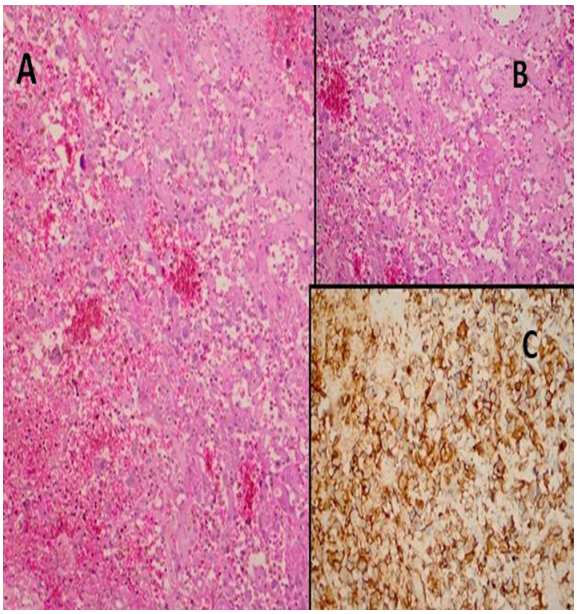

Figure 2. A. Atipical tumoral cells forming vascular structures within large hemorrhagic areas (H&E x100), **B.** Closer look of the tumoral cells; large sytoplasms, aripically appearing cells (H&E x200), C. CD31 positive stained tumoral cells (IHC x200). (H&E : Hematoxylin and Eosine, IHC: Immunohistochemistry).

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tial presentation of the patient, PET CT was performed for general screening purposes. No spread anywhere except the penis was found. The laboratory examinations were also normal. The histopathological evaluation of the incisional biopsy was reported as epitheloid angiosarcoma. On the excisional biopsy, the surgical margines of the mass were intact, however intravascular tumor thrombi were detected. After diagnosis the patient received adjuvant chemotherapy (CTX). In the chemotherapy protocol; Carboplatin dose calculated using AUC 2 was administered for 12 weeks and Paclitaxel was given in a dose of 80mg / m^2 . The patient was followed up with ultrasonograohy and chest X-ray every 3 months. Five years later he admitted to the emergency department with altered mental status and somnolence. On his neurological examinationi decline in the level of consciousness and quadriparesia were detected. On the cranial magnetic resonance imaging (MRI); mass lesions on both parietooccipital areas were observed. The largest one was about 8x30 mm in diameter, with prominent peripheral vasogenic edema, with hemorraghic component, with heterogeneously contrast enhancement (**Figure 1**). Laboratory and other imaging studies were normal in this application. Metastases were not present in other parts of the body. The patient was operated.

Microsurgical technique was used and total resection of the mass at the parietal lobe was performed. The patient had good postsurgical recovery period and was transfered to the oncology department. 300 cGy / Fr total 30 Gy, 2 areas, complex plan, palliative radiotherapy was applied. One month after radiotherapy, the patient died because of general condition deterioration and septicemia.

Pathological findings

The tumors from the penile and cerebral biopsy showed identical morphological and immune staining properties. From the slices prepared of the 2.5x2x2 cm lesion excised at the occipital area; epitheloid-like atipical cells, with large cytoplasms, within large hemorrhagic areas were observed. Immunohistochemically (IHC), the atipic cells showed positive staining with CD31, CD32 and vimentin, and negative CK20 staining. Focal positive staining was observed with PAS staining. Based on the morphological, immunohistochemical findings and the patients clinical history the lesion was reported as metastasis of epitheloid angiosarcoma (**Figure 2**).

DISCUSSION

Angiosarcomas generally constitute less than 1% of all head, face, liver, skin and soft tissue sarcomas.[5] Up to our knowledge almost 30 penile angiosarcomas have been reported in the literature so far.[3] Exposure to vinyl chloride and arsenic, and chronic lymphedema may be accountable as risk factors.[6] Treatment generally consists of radical surgery and adjuvant CTX and RT.[4] In the current literature limited data exist regarding the metastatic behaviour of angiosarcomas. There are few case reports of primary and secondary cerebral angiosarcoma. Metastatic cerebral angiosarcomas are more common compared to primary cerebral angiosarcomas, and generally originate from the right atrium.[5,7] Angiosarcomas metastatizing to the brain are reported to be localized generally in the thorax or abdomen (66%) and cardiac angiosarcomas are reported to metastasize more commonly to the brain.[1,8] In the literature, 2 of 6 splenic angiosarcomas are reported to have metastatize to the brain. Beside being rare, only few cases reporting brain metastasis of penile angiosarcoma are present.[9] Brain metastasis is most frequently found on the parietal (30,7%) and frontal (23%) lobes.[1] The localization of the metastasis in our case also supported these findings. Cerebral metastasis of angiosarcoma is characterized by hemorrhagic transformation with perilesional edema in CT; and on MRI, partial contrast enhancement is present beside hemorrhagia and perilesional edema. It may be confused with cavernous hemangioma due to its hemorrhagic appearance, and with other intracranial tumors due to the peritumoral edema.[5,10,11]

In general, angiosarcomas have very poor prognosis. Tipically, 5 year survival rate is 12%, and metastasis and relaps is seen during the first 2 years.[4] The prognosis may vary due to the primary location of the tumor, surgical resection and tumor size on presentation.[6,12] Particularly in angiosarcomas with brain metastasis the overall survival rate is 2-6 months. In our case; We have achieved better results in terms of survival rate from the literature and life time after metastasis was consistent with the literature.

After the excision of the primary angiosarcoma, CTX and RT may be useful in the prevention of brain metastasis. CTX in cases with brain metastasis is not effective due to the impermeability of the chemotherapeutic agents used in sarcoma treatment.[13] Angiosarcomas have poor prognosis due to their malignant and aggressive course. Our case can also be shown as an example of poor prognosis of angiosarcoma.

Conventionally, sarcoma, renal cell carcinoma and melanomas are accepted as radioresistant tumors. In the past, in sarcoma cases with brain metastasis, primary and fractioned Whole Brain Radiotherapy (WBRT) was used in order to achieve local control, and the average survival was limited to 3 months.[13] Classic treatment methods have been applied in our case and long-term survival after metastasis has not been achieved. Therefore, radiation-resistant brain metastases should be treated more aggressively with stereotactic radiosurgery.

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