CASE REPORT

Metastatic Carcinoma to Vertebra Mimicking as Primary Malignant Vascular Tumour; A unusual Case Report

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Introduction

Papillary intralymphatic angioendothelioma (PILA) or Dabska tumor is an extremely rare vascular tumor. It is discovered by Maria Dabska and only 33 cases have been reported till date [1]. It usually occurs in infants and children although 25 % of the cases occur in adulthood [2]. Most of these occur in the dermis and subcutaneous tissue of extremities [3]. Other locations include spleen, tongue, testis and bone. Intraosseous PILA is very rare and only three cases have been reported. They present as plaques or nodules with asymptomatic growth until it reaches significant size [1, 2]. This lesion present as osteolytic lesion [1, 4, 5]. Surgical excision is the treatment [3, 4]. Histopathology shows interconnecting vascular channels lined by mildly atypical cuboidal endothelial cells with hob nailing and small papillary tufts into lumen [1, 2]. Prognosis is excellent with complete wide excision [1]. We present a case of 42 year old male who came with complains of neck pain of 2 months duration. Lesional biopsy showed metastatic carcinoma with features closely mimicking dabska tumor and which was later confirmed by immunohistochemistry.

Case History

A 42 year old male presented with back pain of 2 months duration. Pain was initially intermittent, dull aching which progressed to be continuous and of shooting type since 1 week. He also complained of weakness in both the upper limbs. There was no history of trauma, fracture or tuberculosis. X ray and magnetic resonance imaging[MRI] of cervical spine showed osteolytic lesion causing complete resorption of C3 vertebral body and compressing the spinal cord anteriorly (Fig. 1a and b). Complete hemogram, urine analysis and liver function tests were normal. Corpectomy with fusion and bone grafting was done and lesional biopsy was sent to histopathology. Histology showed numerous intercommunicating vascular spaces lined by mildly atypical cuboidal cells with hob nailing and papillary tufting (Figs. 1c, d and 2a, b, d). There were also plump slightly larger cells with vesicular nucleus and distinct nucleoli in solid sheets and vague acinar pattern (Fig. 2c). Stroma showed sclerosis and lymphocytes. Adjoining tissue showed cavernous hemangioma like areas. Diagnosis of metastatic poorly differentiated carcinoma was made and advised to evaluate for primary. An alternative differential diagnosis of dabska tumor was also given. Ultrasound and Computed tomography[CT] of abdomen and pelvis was done, which was normal. Prostate specific antigen, carcinoembryonic antigen and alpha feto protein were assessed and were within normal limits. Second opinion of the histopathology from another institute was reported as Angiosarcoma. Hence immunohistochemistry was performed and it showed tumor cells were strongly positive for cytokeratin (Fig. 3a and b), negative for CD 31 (Fig. 3c) and leucocyte common antigen (Fig. 3d). Final diagnosis of metastatic carcinoma with occult primary

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Fig. 1 **a, b** X ray and MRI spine showing osteolytic lesion and complete resorption of vertebra with anterior displacement of spinal cord. **c, d** Histopathology showing numerous vascular spaces with atypical cells with hobnailing and papillary tufting[H&E,X100]

was made. Post operative radiotherapy was given. Patient was followed up subsequently and developed extensive metastatic

lesions within 6 months. The patient was succumbed to death within 8 months.

Fig. 2 **a, b, d** Histopathology showing numerous vascular spaces with atypical cells with hobnailing and papillary tufting[H&E,X100]. **c** Histopathology showing vague acinar pattern with pleomorphic tumour cells[H&E,X100]

Fig. 3 **a, b** Immunohistochemistry showing tumour cells showing CK positivity, **(c)** Leucocyte common antigen negative and **(d)** CD 31 negative

Discussion

Hemangioendothelioma (HE) is rare vascular tumors that have a biologic behavior intermediate between that of hemangioma and angiosarcoma. They account for only 0.5 to 1.0 % of primary malignant bone tumors [4–6]. Papillary intralymphatic angioendothelioma or Dabska tumor was first discovered in 1969 by Maria Dabska [1, 2]. This tumor was termed as malignant endovascular papillary angioendothelioma as two of her cases had lymph node metastases. In 1999, Fanburg-smith JC and his colleagues termed this tumor as PILA, due to its lymphatic origin proved by positive D2-40 and Vascular endothelial growth factor receptor 3. In 2013, World Health Organization (WHO) classification of tumors of soft tissue and bone, this tumor is described as “rarely metastasizing lymphatic vascular neoplasm” [1].

Dabska tumor has a wide age distribution, from birth to 83 years. It usually occurs in dermis and subcutaneous tissues of extremities, especially thigh and buttocks [1–3]. Other locations include spleen, tongue, testis and bone. Intraosseous PILA is very rare and only 3 cases have been reported till date. They presents as a slowly growing asymptomatic cutaneous or soft tissue nodule [1]. In our case, chief complaint was neck pain of 2 months duration.

Intraosseous PILA usually show osteolysis in radiological examination [1]. Unni et al. found a positive correlation between the radiographic picture and the histological grade [4].

High grade tumors have indistinct and irregular margins while low grade tumors have sharp demarcated edges. Cortical destruction often occurs, but periosteal reaction is uncommon. MRI will show intermediate and high signal intensity on T1W and T2W images respectively. CT scan can be used for assessment of cortical destruction and soft tissue penetration. Radiological differential diagnosis includes hemangioendothelioma, metastasis, angiosarcoma and fibrous dysplasia [5]. In our case, X-ray and MRI of cervical spine showed osteolytic lesion causing complete resorption of C3 vertebral body and compressing the spinal cord anteriorly.

Microscopically, PILAs are composed by endovascular papillary proliferations that project into dilated, thin-walled, lymphatic spaces. At high-power view, they demonstrate a pathognomonic hobnail like appearance with scant pink cytoplasm and a prominent nucleus, with little or no cytological atypia. Mitotic figures are rare. Endothelial cells are immunoreactive for CD34, CD31 and factor 8 related antigen [2]. CD31 has high sensitivity and specificity for vascular endothelial cells [1]. In our case, histology showed features of dabska tumour. There were also plump slightly larger cells with vesicular nucleus and distinct nucleoli in solid sheets and vague acinar pattern. Adjoining tissue showed cavernous hemangioma like areas. The histopathological differential diagnosis includes epithelioid hemangioendothelioma, retiform hemangioendothelioma, angiosarcoma and metastatic carcinoma. Epithelioid hemangioendothelioma is mainly located

in long tubular bones of the lower extremities and pelvis, less frequently in the vertebral column, upper extremity, and flat bones. It usually occurs in 4th–5th decade of life and is often multicentric [5]. Histology shows epithelioid cells streaming off medium to large sized vessels, usually veins, arranged in strands, cords and solid nests. Mild atypia and occasional mitosis may be seen. The cells have abundant hyalinised cytoplasm with lumen containing erythrocytes which represent abortive vascular lumina [6, 7].

Retiform hemangioendothelioma is a distinctive form of low grade angiosarcoma, most commonly occurring in extremities of young adults. Histology shows proliferation of arborizing blood vessels arranged in retiform pattern, resembling rete testis. Stroma usually has prominent lymphocytic infiltrate. Some authors consider this tumor as adult counterpart of dabska tumor and coined the term hobnail hemangioendothelioma to encompass both neoplasms [8].

Angiosarcoma is a malignant vascular neoplasm with high incidence of local recurrence, metastasis and high mortality rate. Histologically there is significant cytologic atypia, presence of conspicuous mitotic figures, tumor dissecting collagen bundles and hemorrhagic background [7, 8].

Metastatic carcinomas have more pleomorphic, mitotically active tumor cells evoking desmoplastic response [7]. As most common malignant bone tumor is metastasis, vertebra being the commonest site, and histology showing vague acinar pattern on deeper tissues, we gave a diagnosis of metastatic poorly differentiated carcinoma and suggested immunohistochemistry. Search for primary by radiological examination and by tumor markers did not fetch any positive results. Tumor cells were strongly positive for cytokeratin, negative for CD 31 and leukocyte common antigen. Hence the final diagnosis of metastatic carcinoma with occult primary was made. Patient was followed up subsequently. Due to extensive metastatic lesions and co morbid conditions patient was succumbed to death within 8 months.

Treatment is determined by the grade, location and spread of the tumor. Wide surgical resection is recommended. Radiotherapy is indicated for a widespread or irresectable tumor to prevent tumor progression. Role of chemotherapy is not well established [3–5]. In our case, corpectomy with fusion and bone grafting was done. Since resection was not possible in our case, post operative local radiotherapy was given.

Conclusion

The metastasis is the most common malignant bone tumor and it usually affects thoracic and lumbar vertebra. But high cervical vertebral metastasis is uncommon. Histology in our case had all classical features of PILA. This finding is very rare and is very much misleading for primary microscopic diagnosis, especially when it occurs at a site where repeat biopsy or wide excision is not possible. Differential diagnosis should be kept in mind and discussed with the surgeons, as both of our motives are correct diagnosis and complete cure of the patients.

Compliance with Ethical Standards

Source of Support nil

Conflict of Interest None

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