

# A Case Report of Aggressive Metastatic Parachordoma

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To the Editor;

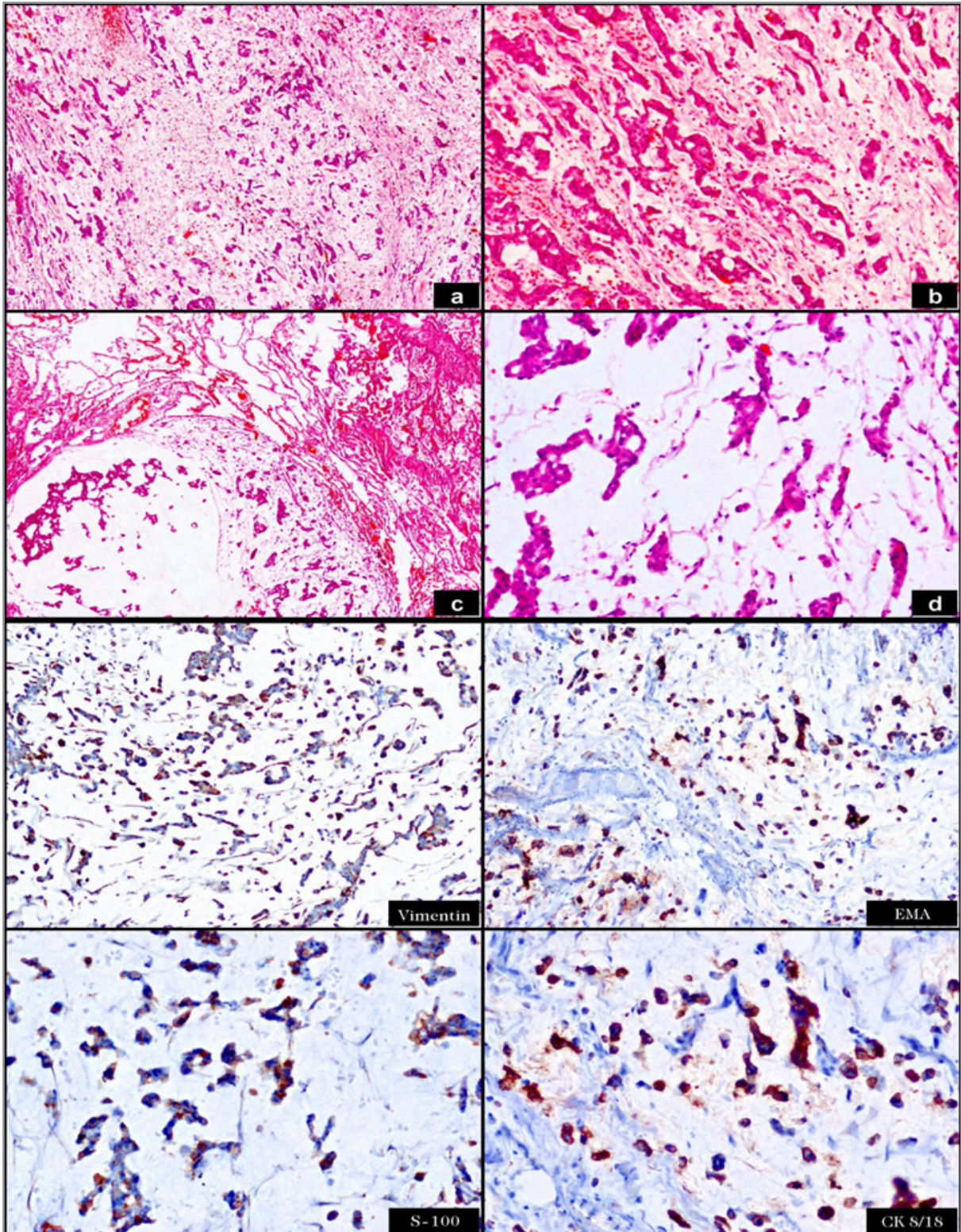
Parachordoma, first described by Laskowski in 1951, is a rare tumor of soft tissue. Few than 100 cases have been reported in the literature. In 1977 Dabska M. collected 10 cases of parachordoma and he described it as a slow growing, locally invasive tumor.<sup>1</sup> Since then, metastatic cases have been described but they are also so rare. In this report, we present a case of parachordoma with an aggressive nature, rapidly metastasizing to bone, lung and intraabdominal organs.

A 49 year-old male presented with a painful right inguinal mass progressively enlarging in the previous 2 months. Physical examination showed a 10 centimeters mass in groin extending towards symphysis pubis. The excisional biopsy was performed. All tissue samples were fixed in 10% neutral-buffered formalin, processed routinely, and embedded in paraffin. Then, standard 4 micrometer-thick sections were obtained from all blocks and stained with H&E for the light microscopic evaluation. Furthermore, sections from representative blocks were pretreated by heat for antigen retrieval and stained against vimentin, EMA, S-100 (Novocastra, 1:100), and CK 8/18 (Santa Cruz Biotech, 1:150) primary antibodies. The tumor was unencapsulated but well-circumscribed macroscopically. Its cut-surface was glistening and had a rather fragile consistency. Microscopy revealed that the tumor was composed of chords and chains of the relatively uniform, eosinophilic cells in a myxoma-like stroma. Immunohistochemically, the neoplastic cells

were highlighted by vimentin, EMA, S-100 protein, and CK 8/18 (Figure 1). Based on the clinical, morphological and the immunophenotypical features, a diagnosis of parachordoma was made.

The patient was followed without an adjuvant therapy. Four months after resection, the patient relapsed with a mass in the right spermatic cord. With ultrasonography a lobulated solid lesion was noted in right spermatic cord. Tru-cut biopsy was consistent with parachordoma. Right spermatic cord excision and right orchiectomy were performed. The histopathologic and immunohistochemical examination confirmed the diagnosis of parachordoma. Two months after the second operation, the patient presented again with right hip pain. Magnetic resonance imaging showed a lesion on the head of the right femur. Due to absence of metastasis in other parts of the body, palliative radiotherapy was the only modality for tumor control. Pain improved with radiation.

At the 10th month of follow up, a 6x5 cm mass in the inferior lobe of left lung and 4x3 cm lesion in the right hilum were encountered. Positron emission tomography demonstrated pathological uptake at mediastinal lymph nodes, left ischium, in a nodule in the right inferior lobe of the lung and left pleural effusion. Other parts of the body were free of metabolic activity. Bronchoscopic biopsy confirmed the diagnosis of parachordoma. The tumor was resected grossly including the pericardial invasion.



**Figure 1.** The figure shows the histopathological and the immunophenotypical features of the tumor. Both the primary site, which was right inguinal region (a and b), and the pulmonary metastases (c and d) look alike histologically. Bland and uniform-looking eosinophilic cells form chords, chains and small islands in fairly loose interstitial matrix reminiscent myxomatous tumors. Neoplastic cells express vimentin, epithelial membrane antigen (EMA), S-100 protein and cytokeratin 8/18 (bottom-two rows).



Two weeks after the operation patient started to complain about his back and right hemithorax pain which was refractory to narcotic analgesics. A new computerized tomography showed, 18x12 mm lesion in the liver, intraabdominal lymph nodes and 7.7x5.5 cm lesion in abdomen infiltrating right adrenal gland and aorta. The rapidly progressing parachordoma was treated with adriamycin 20 mg/m<sup>2</sup>/week with palliative intent. After 6 courses of therapy, symptoms improved but disease progressed radiologically. Also during this chemotherapy period the patient had a thoracal imaging with MRI for his back pain. An expansile lesion on T4 spinal process was determined and local radiotherapy was applied.

The patient was followed with palliative radiotherapy and narcotic analgesics. The progression under adriamycin therapy conveyed the treatment modality to imatinib mesylate (800 mg/day P.O.) which was continued for 4 weeks. Under palliation and Imatinib therapy, the patient progressively deteriorated and cholestasis ensued. The rapidly progressing tumor was refractory to palliative management such as external biliary drainage. The patient died as result of hepatic and cardiovascular insufficiency.

Although its histological origin is unknown, there are various theories that parachordomas are related with chordomas, extraskelatal myxoid chondrosarcomas and myoepitheliomas. World Health Organization has classified parachordomas in the same class as mixed tumors and myoepitheliomas in 2002.<sup>2</sup> However parachordoma is thought to be a possible low grade sarcoma because of the small number of cases with only limited follow up information.

Histopathologically, parachordomas are circumscribed but unencapsulated masses that were composed of cords and chains of polygonal, eosinophilic cells in a myxoid stroma. Tumor cells are predominantly rounded eosinophilic cells arranged in short cords and chains. Some tumor cells are multivacuolated like physaliferous cells. Nuclear atypia varies from mild to moderate and mitoses are rare. Immunohistochemically; parachordomas are positive for CK 8/18, epithelial membrane antigen (EMA), S-100 protein and vimentin. Unlike chor-

domas, parachordomas are negative for CK 1/10. Also parachordomas show a nestlike pattern of type 4 collagen. Chordomas are strongly positive for CK 8/18, CK 1/10 and vimentin. Like mentioned above two tumors can be distinguished by their CK 1/10 affinity. In differential diagnosis extraskelatal myxoid chondrosarcomas are negative for all cytokeratins.<sup>3,4</sup> In our case the tumor was composed of chords and chains of the relatively uniform, eosinophilic cells in a myxoma-like stroma and the neoplastic cells were highlighted by vimentin, EMA, S-100 protein, and CK 8/18.

The analysis of the cases reported in literature shows that parachordoma is generally a pathology of younger population, mostly under 30 and more commonly occurs in male patients. Extremities are the most commonly reported tumor site. Rarely gastric mucosa, chest wall, cranium and retroperitoneum have been described as primary sites. Usually an indolent nature, slow growth and occasional late recurrence have been reported. Its metastatic potential is poorly defined and there are few than 10 metastatic cases in the literature.<sup>5-10</sup> Recurrent and metastatic presentation differs in these few cases and time to progression takes months to years. Most metastatic patients are adults over 40 year of age. The experienced metastatic sites are usually lung, bone, brain, subcutaneous tissue and regional lymph nodes. Clinical approach generally involves palliative maneuvers.

Our patients features were similar with the literature experience of metastatic cases regarding age and clinical aggressivity. However hepatic and intraabdominal metastasis observed in our patient have not been reported previously. Intraabdominal recurrence within months after surgical approach and refractory nature to cytotoxic therapy was challenging.

Systemic treatment experience is limited in metastatic cases and the results are not encouraging. Guedes et al reported refractoriness to ifosfamide and etoposide combination.<sup>9</sup> Abe et al. reported ineffective adriamycin based combination therapy in metastatic parachordoma.<sup>7</sup> Gemcitabine-docetaxel combination experience in neoadjuvant setting was again useless.<sup>8</sup> The subjective response couldn't be supported with radiological imaging in our case

and further clinical deterioration changed the therapy. A case of adjuvant imatinib following lung metastasis resection was reported previously<sup>10</sup> but our patient progressed rapidly under imatinib treatment and died in 5 weeks.

In summary, although parachordoma is generally expected as an indolent and rarely metastatic neoplasm, the close follow up after surgery is crucial. No active systemic treatment exist for metastatic disease. Periodical lung and bone imaging may help to detect early recurrences and provide early surgical intervention. To clarify the nature of this rare pathology, further experience with large number of cases is needed.

#### REFERENCES

1. Dabska M. Parachordoma: a new clinicopathologic entity. *Cancer* 40: 1586-1592, 1977.
2. Fletcher C, K. Unni, F. Mertens. World Health Organization Classification of Tumours: Pathology and Genetics of Tumors of Soft Tissue and Bone. 2002: Lyon: IARC Press.
3. Folpe AL, Agoff SN, Willis J, Weiss SW. Parachordoma is immunohistochemically and cytogenetically distinct from axial chordoma and extraskelatal myxoid chondrosarcoma. *Am J Surg Pathol* 23: 1059-1067, 1999.
4. Fisher C, Miettinen M. Parachordoma: a clinicopathologic and immunohistochemical study of four cases of an unusual soft tissue neoplasm. *Ann Diagn Pathol* 1: 3-10, 1997.
5. Miettinen M, Gannon FH, Lackman R. Chordomalike soft tissue sarcoma in the leg: a light and electron microscopic and immunohistochemical study. *Ultrastruct Pathol* 16: 577-86, 1992.
6. Kinoshita G, Yasoshima H. Fatal parachordoma. *J Orthop Sci* 12: 101-106, 2007.
7. Abe S, Imamura T, Harasawa A, et al. Parachordoma with multiple metastases. *J Comput Assist Tomogr* 27: 634-638, 2003.
8. De Comas AM, Deavers MT, Raymond AK, et al. Intra-neural parachordoma of the arm with regional metastases. *Skeletal Radiol* 40: 943-946, 2011.
9. Guedes A, Barreto BG, Barreto LG, et al. Metastatic parachordoma. *J Cutan Pathol* 36: 270-273, 2009.
10. Lococo F, Cesario A, Meacci E, et al. Pulmonary metastases from parachordoma. *Ann Thorac Surg* 88: e9-10, 2009.

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