ABDOMINAL PARACHORDOMA – A CASE REPORT

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Summary

We report a very rare case of abdominal parachordoma. Parachordoma is indolent in nature, grows slowly and it has occasional recurrence after 3 months to 12 years and rare metastases.

A 70-year-old female presented to our Department complaining of abdominal pain. Clinical examination showed a tumor mass in the pelvis and abdomen. Surgery was indicated. Histopathological and immunohistochemical analysis of resected tumor tissue confirmed the diagnosis of parachordoma with metastases in the sigma and omentum.

KEY WORDS: *parachordoma, abdomen, metastases*

ABDOMINALNI PARAKORDOM – PRIKAZ SLUČAJA

Sažetak

Prikazujemo vrlo rijedak slučaj abdominalnog parakordoma. Parakordom raste sporo, rijetko metastazira, a recidiv se može pojaviti nakon 3 mjeseca do 12 godina.

Sedamdesetogodišnja pacijentica primljena je na Odjel za ginekološko-onkološku kirurgiju Klinike za tumore KBC „Sestre milosrdnice" zbog bolova u donjem dijelu trbuha. Kliničkom obradom dijagnosticirana je tumorska masa u zdjelici i trbuhu. Učinjen je kirurški zahvat. Histološka i imunohistokemijska analiza potvrdila je dijagnozu parakordoma s metastazama u sigmi i omentumu.

KLJUČNE RIJEČI: *parakordom, abdomen, metastaze*

INTRODUCTION

Parachordoma is a very rare peripheral soft tissue tumor of unknown origin, with fewer than 50 cases described in the English literature (1). The tumor initially mentioned as "chordoma peripericum" by Laskowski in 1951, was later renamed to parachordoma by Dabska (2) who reported initial 10 cases in 1977. The tumor is usually circumscribed and firm with a variety of histological patterns which to some extent resembles chordoma, but with a wider range of clinical appearance and specific cytokeratin immunophenotype (3). This tumor that typically occurs in the fourth decade of life is predominantly situated in the lower extremities and almost equally distrib-

uted in men and women. Parachordoma is indolent in nature, grows slowly and it has occasional recurrence after 3 months to 12 years and rare metastases with 5 fatal outcomes reported so far (4). The surgical management usually includes wide resection.

PATIENT REPORT

A 70-year-old female patient presented with low abdominal pain of a 6-month duration. The patient had no other symptoms. Physical examination revealed a palpable mass in the lower abdomen. Tumor markers were within the limits of normal. Ultrasound and computed tomography detected a solid mass in the pelvis and abdomen. Surgical treatment was indicated. Intraoperatively, we found a large tumor located in the lower abdomen with multiple adhesions to the colon. After careful detachment from the colon, the tumor was found to have two nodules on the sigma, one of 5 mm and the other of 11 mm in diameter. The tumors were removed entirely. The omentum was consumed by multiple tumor nodules. Total omentectomy was performed. The uterus was closely attached to the tumor mass, and after adhesiolysis, a total abdominal hysterectomy with bilateral salpingo-ovariectomy was performed.

The resected larger tumor measured 30x15 cm. Serial slicing revealed extensive central necrosis. Microscopically, the tumors consisted of epitheloid polygonal cells with eosinophilic, variably vacuolated cytoplasm. Nuclear mitotic figures were rare (1-3 on 10HPF). Immunohistochemical analysis confirmed the diagnosis of parachordoma. All tumor cells showed positive reactions for vimentin, cytokeratin (CK) 8/18, S-100 protein, epithelial membrane antigen (EMA), type IV collagen and CD 99 as well as positive focal reactions for NSE and negative reactions for CK 1/10, 19, 20.

She checks up regularly. To this date, 12 months after surgery, there has not been any evidence of local or distant recurrence.

DISCUSSION

Parachordoma grows slowly, tends to have a local destructive nature and metastasizes rarely (six cases described so far). It was first reported in 1951 as "chordoma periphericum" by Laskowski and it was later re-described and renamed to "parachordoma" by Dabska (2). Differential diagnosis often includes chordoma and extraskeletal myxoid chondrosarcoma (1) which was considered in the pathological specimens in this case. Unlike chordoma, the parachordoma reactivity to type IV collagen is much stronger as presented in our case as well. Chordoma usually shows CK 1/10 and CK 19 that were both negative in the present immunohistochemical analysis. Extraskeletal myxoid chondrosarcoma was excluded since the epithelial markers EMA and CK 8/18 were positive. Since its first description, and in fewer than 50 cases reported (1), the tumor was mainly located in the lower (51.2%) and upper extremities (26.8%). The origin of parachordoma is still uncertain; in the literature (5) it has been suggested to be of ectopic nests of notochord, Schwann or neuron-related cells, specialized synovial cells and totipotent mesenchymal cells.

In our case, the tumor had unusual multiple local metastases on the sigma and omental surface. There have been six cases of metastases from parachordoma (6-11). Two of them (6, 7) belonged, according to the pathological finding, more to the chordoma tumor origin than to the parachordoma. Limon et al. (8) reported a case with lymph node metastases in the chest wall, similar to a recent report by Lococo et al. (9) who reported pulmonary metastases after a three-month free period following local surgery of the left hip due to primary parachordoma. Abe et al. (10) presented a case of multiple metastases of the parachordoma primarily located in the calf. The patient died 32 months after surgery despite radiotherapy and chemotherapy. Another metastatic behavior of calf primary parachordoma was noted in the 60-year-old male who died after 4 months following the surgical treatment due to lung and brain metastases (11).

CONCLUSION

Our patient report adds a new insight into the possible location of parachordoma and its aggressive behavior presented with metastases in surrounding tissue, i.e. the sigma and omentum. Parachordoma should be considered as a locally

aggressive, low grade tumor with a possibility of metastatic development.

REFERENCES

1. Clabeaux J, Hojnowski L, Valente A, Damron TA. Parachordoma of soft tissue of the arm. Clin Orthop Relat Res 2008; 466: 1251-6.
2. Dabska M. Parachordoma: a new clinicopathologic entity. Cancer 1977; 40: 1586-92.
3. Fisher C. Parachordoma exists-but what is it? Adv Anat Pathol 2000;7:141-8.
4. Imlay So, Argnyi ZB, Stone MS et al. Cutaneous parachordoma. A light microscopic and immunohistological report of two cases and review of literature. J Cutan Pathol 1998; 25: 279-84.
5. Cho SH, Sung NK, Jung KJ, Lee YH, Park YC, Kim HK, Park SY, Park KS, Ko SM. Parachordoma of the chest wall: case report. J Korean Radiol Soc 2004; 51: 295-8.
6. Miettinen M, Gannon FH, Lackman R. Chordoma like soft tissue sarcoma in the leg: a light and electron microscopic and immunohistochemical study. Ultrastruct Pathol 1992; 16: 577–86.
7. Carstens PH. Chordoid tumor: a light, electron microscopic, and immunohistochemical study. Ultrastruct Pathol 1995; 19: 291–5.
8. Limon J, Babinska M, Denis A, Rys J, Niezabitowski A. Parachordoma: a rare sarcoma with clonal chromosomal changes. Cancer Genet Cytogenet 1998; 102: 78–80.
9. Lococo F, Cesario A, Meacci E, Cusumano G, Margaritora S. Pulmonary metastases from parachordoma. Ann Thorac Surg 2009; 88: 9-10.
10. Abe S, Imamura T, Harasawa A, Ishida T, Unno K, Tateishi A, Tokizaki T, Yorikawa J, Matsushita T. Parachordoma with multiple metastases. J Comput Assist Tomogr 2003; 27: 634–8.
11. Kinoshita G, Yasoshima H. Case report: fatal parachordoma. J Orthop Sci 2007; 12: 101–6.

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