Metastatic chondroblastoma

Errol U. Hutagalung*, Saukani Gumay[f]

Abstrak

Dilaporkan suatu kasus kondroblastoma yang merupakan tumor jinak tulang dengan metastasis ke paru-paru dan dinding abdomen. Perjalanan penyakit bersifat progresif seperti suatu proses keganasan. ***(Med J Indones 2001; 10:57-9)***

Abstract

A case of chondroblastoma, which is a benign tumor of the bone, with distant metastases to the lung and abdominal wall is reported. The clinical course of the disease was progressive like that of malignant process. ***(Med J Indones 2001; 10:57-9)***

Keywords *: chondroblastoma, bone tumor*

Chondroblastoma is an uncommon benign bone tumor having special radiologic and histologic features.[1-3]

Chondroblastoma was first described by Codman in 1931 as epiphyseal chondromatous giant cell tumor (GCT), a variant of GCT of the bone. Only in 1942 did Lichtenstein and Jaffe established the tumor as a distinct entity different from GCT and described it as a benign chondroblastoma.[4,5] Despite its benign character and benign histologic feature, the tumor is known to have distant metastases.

Literature study as recent as 1998 revealed only 12 cases of chondroblastoma with distant metastasis.[16] Herewith we report a case with metastases to the lungs and abdominal wall, which is the first to be reported from Indonesia.

CASE REPORT

The patient, a 16-year-old Indonesian female, was admitted to the hospital complaining of mass on the right knee since one month. History of accidents and fever were denied. A week after the mass was recognized, another mass appeared on the abdominal wall below the navel, with the size of a marble and gradually enlarged.

On physical examination, the patient seemed to be in good condition. Local examination of the right knee revealed a mass measuring 9 x 7 x 3 cm, firm in consistency, warmer than the surroundings, and painful on pressure. Movement of the right knee appeared to be limited. On the abdominal wall, under the navel, was another firm mass, measuring 3 x 2.5 x 2 cm, and not painful on pressure.

Laboratory findings were within normal limits, except for a slight increase in serum alkaline phosphatase. Chest X-ray indicated no abnormalities. X-ray of the right knee revealed an osteolytic lesion with sclerotic borders on the proximal tibia, at the region of the meta-epiphysis.

Based on the clinical and radiologic findings the possibility of osteosarcoma was considered. An incisional biopsy was performed on the tumor, and pathological examination indicated a chondroblastoma, without any signs of malignancy. Based on the pathology, curettage was performed and the defect was filled with autologous bone graft taken from the iliac crest. The tissue obtained from the curettage was sent for pathologic examination, which indicated no signs of malignancy, and showing chicken wire ossification which is a unique feature of chondroblastoma.

* *Subdivision of Orthopaedics, Department of Surgery, Faculty of Medicine, University of Indonesia and Dr. Cipto Mangunkusumo General Hospital, Jakarta, Indonesia*

[f] *Department of Anatomic Pathology, University of Indonesia School of Medicine and Dr. Cipto Mangunkusumo General Hospital, Jakarta*

Three weeks after surgery the patient showed shortness of breath, which appeared to be due to a left pleural effusion and bilateral bronchopneumonia. Puncture of the effusion was performed, from which about 1000 mL of serohemorrhagic fluid flowed out. Cytologic examination of the fluid did not show any indication of malignancy. The patients' breathing returned to normal after the puncture.

A week later she became short of breath again, apparently due to reaccumulation of fluid in the left pleural cavity. It was decided to install a Water Sealed Drainage (WSD), which delivered about 800 mL of serohemorrhagic fluid. Cytologic examination still did not reveal any malignancy.

Meanwhile, the mass on the abdominal wall enlarged and it was decided to perform an excisional biopsy under general anesthesia. The excised tissue was sent for pathologic examination, which revealed similar histologic features as the pervious ones, confirming a chondroblastoma.

Three days afterwards another effusion developed in the right pleural cavity, which upon puncture extruded about 600 mL of serohemorrhagic fluid. The diagnosis was then established as aggressive chondroblastoma with metastases to the lungs and abdominal wall.

The disease progression is clinically a malignant process as indicated by decrease in body weight of 7 kg within a period of one and a half months, development of anemia, bilateral pleural effusion and enlargement of a metastatic mass in the abdominal wall, as well as rapid enlargement of the lesion on the right knee. It was therefore decided to administer chemotherapy and pleurodesis.

Three days after giving the first cycle of cytostatic, pleurodesis was performed with about 20 mL of terramycin. While undergoing pleurodesis the patient suddenly appeared to be in pain, developed neurogenic shock and died. Autopsy was not performed.

DISCUSSION

Chondroblastoma is a solitary benign tumor of the bone which is most frequent in the second decade (10 - 20 years) and having unique radiologic and histologic features. As revealed on X-ray, the tumor is located at the epiphysis and appeared to have a relatively round shape, showing distinct borders and thinning and erosion of the surrounding area.[7]

By definition a benign tumor does not show the ability of spreading to other sites (metastases). However, a study of the literature revealed a number of cases of chondroblastoma reported as having metastases, which on histopathologic examination showed benign features similar to those of the original tumor. Therefore the term aggressive or malignant chondroblastoma was considered inappropriate by some, as the term was related to its clinical development and not its histopathologic description.[1]

Metastases were generally to the lungs, but it had also been reported into various organs such as the talus, ribs, abdominal viscera, and other soft tissues.[9,10] Metastases appeared to develop following a surgical procedure such as curettage, and was reported to be within a period of 5 months - 33 years post-operatively.[5] In the case reported by Khalili et al,[10] multiple metastases occured at different times within 8 - 17 years; all of them into soft tissues, whereas the lungs remained clear. Pathologic examination of the metastatic tissues all showed the same benign features of the origical tumor.

It was believed that metastases developed by means of emboli of tumor cells as a result of surgical manipulation or curettage.[10] This theory could not explain the metastases in the case reported by Kyriakos et al,[5] where lung metastases were already observed prior to surgery. Similarly, in the present case, metastases were already apparent in soft tissues / abdominal wall prior to surgery.

The clinical, radiological and histopathologic findings in the present case correspond to those reported in the literature. The tumor develop progressively, similar to that described by Kyriakos et al.[5] It was reported that the lung metastases and local recurrence at the site of the original lesion following curettage developed progressively, leading to the decision to give chemotherapy of vincristin, cyclophosphamide and adriamycin, which turned out to be of no benefit.[5]

In the present case, the tumor also developed progressively as indicated by lung metastases with the manifestation of productive pleural effusion, and signs of local recurrence that rapidly enlarged within 3 weeks post-operatively. It was decided to give chemotherapy with doxorubicin, vincristin and cyclophosphamide, albeit only for one cycle as the

patient died of neurogenic shock while undergoing pleurodesis.

Of the cases reported with recurring and multiple distant metastases, the patients apparently survived 5, 6, 17 and 30 years after the diagnoses was established.[6,9-11] The case reported by Kyriakos et al,[5] in which the tumor showed progressive development, died 5 years after diagnosis. The case described by Wirman et al[12] had a relatively slow development, the patient refused to undergo repeated surgery for resection of the recurring tumor, and later died after 34 years. The progression of the tumor reported by Wirman et al could be considered as representative of the natural history of chondroblastoma which do not receive treatment.

The present case ended with the death of the patient only 9 weeks after being diagnosed, however, the death was not on account of the tumor itself but due to the excessive pain and neurogenic shock while undergoing pleurodesis.

REFERENCES

1. Unni KK. Dahlin's Bone Tumors. General Aspect and Data on 11.087 Cases. 5th ed. New York: Lippincott-Raven, 1996: 47 - 57.
2. Turcotte RE, Kurt AM, Sim FH, Unni KK, McLeod RA. Chondroblastoma. Hum Pathol 1993; 24: 944 - 9.
3. Fechner RE, Mills SE. Atlas of Tumor Pathology. Tumor of the Bones & Joints. Washington: Armed Forces Institute of Pathology, 1992: 91 - 5.
4. Linchtenstein L. Bone Tumors. 4th ed. St. Louis: Mosby, 1972: 45 - 55
5. Kyriakos M, Land VJ, Penning HL, Parker SE. Metasttic chondroblastoma. Report of a fatal case with review of the literature on atypical, aggresive and malignant chondroblastoma. Cancer 1985; 55: 1770 - 89.
6. Jambhikar NA, Desai PB, Chitale DA, Patil P, Arya S. Benign metastasizing chondroblastoma. A case report. Cancer 1998; 82: 675 - 8.
7. Hudson TM, Hawkins IF. Radiological evaluation of chondroblastoma. Radiology 1981; 139: 1 - 10.
8. Mirra MJ, Bone Tumors: Diagnosis and Treatment. Toronto: Lippincott, 1980: 219 - 33.
9. Birch PJ, Buchanan R, Golding P, Pringle JAS. A case report. Chondroblastoma of the rib with widespread bone metastases. Histopathology 1994; 25: 583 - 5.
10. Khalili K, White LM, Kandel RA, Wunder JS. Chondroblastoma with multiple distant soft tissue metastases. Case report. Skeletal Radiol 1997; 26: 493 - 6.
11. Riddel RJ, Louis CJ, Bromberger NA. Pulmonary metastases from chondroblastoma of the tibia. Report of a case. J Bone Joint Surg 1973; 44B: 848 - 53.
12. Wirman JA, Crissman JD, Aron BE. Metastatic chondroblastoma. Report of an unusual case treated with radiotherapy. Cancer 1979; 44: 87 - 93.