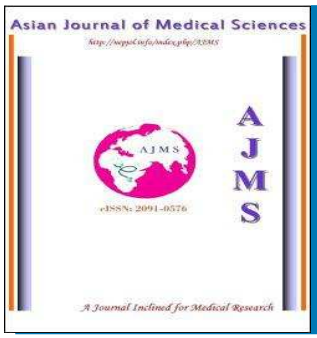

SACRAL CHORDOMA METASTATIC TO BILATERAL CLAVICLE- A RARE CLINICAL PRESENTATION AND REVIEW OF LITERATURE

CASE REPORT AND REVIEW OF LITERATURE, Vol-4 No.4

Asian Journal of Medical Science, Volume-4(2013) http://nepjol.info/index.php/AJMS

[1]Rashi Agrawal, [2]Sudarsan De, [3]Alpana Srivastava, [4]Sweety Gupta, [5]Dinesh Singh, [6]Sandeep Agrawal. [1,2,4,5,6] Department of Radiation Oncology ,Galaxy Cancer Institute , Pushpanjali Crosslay Hospital, India [3]Department of Pathology, Pushpanjali Crosslay Hospital, India.

ABSTRACT

Chordoma is an uncommon tumor , so the metastatic cases generally are given more attention. Various cases with central nervous system involvement, cutaneous and mandible metastases have been reported. Metastases to bone is noted in literature but metastases to bilateral clavicle is not reported yet to the best of our knowledge. We hereby present a rare case of sacral chordoma with clavicular metastases.

Key Words- Metastasis, Chordoma, Clavicle.

CORRESPONDENCE:

Dr Rashi Agrawal
J 48
Patel Nagar First
Ghaziabad
Uttar Pradesh, India
201001
(M) +919891483550
Email: drrashi.ag@gmail.com

"Though metastases to bone are noted in literature but metastasis to clavicle from sacral chordoma is not reported to the best of our knowledge. Here we are presenting data regarding clavicle metastasis from primary sites other than chordoma, patterns of metastasis in chordoma and a rare presentation of chordoma."

INTRODUCTION

Chordoma is a rare ,slow growing neoplasm and known for local recurrence .It has modest propensity for distant metastasis but not an uncommon occurrence. The main metastatic sites are lymph nodes, liver, lung, skin and bones. Here we present a case of primary sacral chordoma with metastases to medial end of both clavicles and systemic spread.

CASE STUDY

A 40 years old gentleman presented in October 2005 with progressively increasing swelling in the lower back. On clinical examination there was 6x5 cm, hard ,fixed and nontender swelling in the sacral region. He was investigated and diagnosed as primary sacral chordoma and underwent surgery. He remained asymptomatic for fifteen months. In February 2007, the disease recurred at the same site. The disease was unresectable so was considered for surgical debulking of the tumor followed by adjuvant radiotherapy at another hospital in March 2007. He was asymptomatic for three years when he presented at our insitute on 6.02.2010 with complaint of swelling and pain over right upper chest for last four months and pain in right arm for last fifteen days. On clinical examination at the medial end of right clavicle a 10x8 cm and at the left clavicle 2x2 cm size swelling firm to cystic in consistency, nontender ,fixed to underlying structure, were present (Figure 1). On left scalp region 5x5 cm swelling with similar characteristics was present. Systemic examination was unremarkable. Trucut biopsy from clavicular swelling was done on 06.02.2010 and reported as malignant large cell tumor consistent with metastatic chordoma (Figure 2). Mitotic figures were frequent. Large polygonal cells with large pleomorphic nuclei were found. Contrast enhanced CT scan chest was done on 01.02.2010 that revealed expansile destructive involving medial end of right clavicle of about 8x8cm size with compression of underlying veins and dilated veins in right axilla. Another lesion of 2x2 cm with same characteristics was present at the medial end of left clavicle. Multiple rounded metastatic nodules were present in both lungs and liver. He was considered for palliative radiotherapy to painful region. He received 20 Gy in five fractions by direct anterior field to clavicular region by 6 MV photons. Now he is in regular follow up and is free of pain till date.

Figure 1- Hard, fixed swellings on medial end of both clavicles

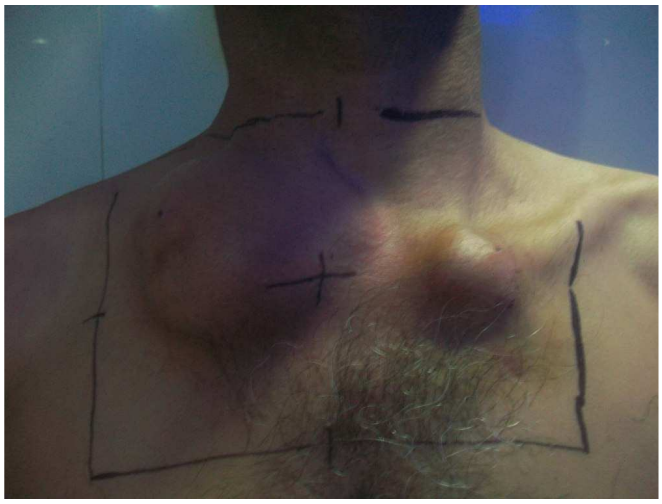

Figure:2 Biopsy from Clavicle showing large to polygonal cells with extracellular mucin like material (H& E,40X)

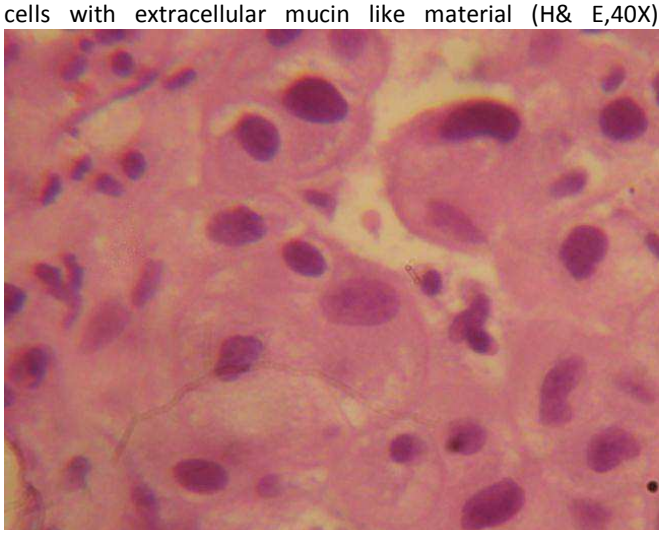

Table 1- Percentage of metastasis from chordoma

Study	Total patient	% of metastasis
Fagundes et al[6]	204	6.3%
Sundareson et al[7]	18(vertebral) 36(sacral)	60% 27%
Volpe et al 1983	25(spinal)	2%
Dahlin et al[4]	59	10%
Higinbotham et al[5]	**46**	**48%**

Table 2- Clavicle metastasis from different primary sites

Author	Total patient	No of clavicle metastasis	Primary site
Lynn et al1986	38	5	Malignant pheochromocytoma
Bose et al 1998	1	1	Thyroid carcinoma
Simon et al1986	1	1	Ca Bronchus
Pittas et al 2000	146	5	Ca thyroid
Sengelov et al 1995	85	2	Ca urinary bladder
Althausen et al 1997	38	1	Renal cell carcinoma
Talbot et al 1989	**48**	**1**	**Ca large intestine**

DISCUSSION

Chordomas are rare, aggressive and slow growing lesions and account for approximately 1-4 % of all primary malignant bone tumors.[1] Approximately 50% chordomas arise in the sacrococcygeal region. Majority of these occur between the ages of 40-70 years (average 55 years).These occur almost twice as frequently in men compared to women. Classical radiological findings of chordomas are those of a bony lytic lesion with an accompanying soft tissue mass and intratumoral calcification. The osteolysis is well circumscribed without an osteosclerotic rim, but sometimes associated with bulging of the cortex. A solid tumor with cystic areas is seen in approximately 50% of cases.[2,3]Histologically there may be some variability, but the major features include a lobular cellular structure, arrangements in cords or clusters, an ample intercellular mucinous matrix, and large physaliphorous cells. Primary treatment for chordomas is wide resection. Metastasis is usually a late event and most often occurs in young patients those with sacrococcygeal or vertebral tumors and those with atypical histological features.

Chordomas metastasize in 3 to 48% of the patients harboring these lesions. In literature there is much discrepancy regarding data of metastasis in chordoma. According to Dahlin and MacCarty the 10% incidence of metastasis often cited in literature may be misleading, since many reporters have failed to give reasonable proof that the neoplasms were chordomas.[4] In sharp contrast is the overall 48 percent incidence of distant metastases reported by Higinbotham and coworkers in their 46 cases.[5] Here we are presenting a table regarding data of metastasis from chordoma.(Table 1)

In descending order of metastasis, the organs involved by metastasis include the lungs, lymphnodes, liver, bone, skin, muscle, peritoneum, heart, pleura, retroperitoneum, spleen, duramater, kidney and adrenal glands.[6] As chordoma is rare tumor, the metastatic cases generally have

more attention and been given rise to more publications.

After reviewing the literature in 1979 chambers et al reported 40 chordoma patients with lung metastasis, 23 with lymphnode, 15 with liver and 12 with bone metastasis.[8]

Metastasis to clavicle is well known in carcinomas of thyroid, urinary bladder, breast, kidney,colon and also from malignant pheochromocytoma among others. Bilateral symmetrical metastases to clavicular heads from bronchiogenic carcinoma is also reported.[9]

Though metastases to bone is noted in literature but metastasis to clavicle from sacral chordoma is not reported to the best of our knowledge. Here we are presenting data regarding clavicle metastasis from primary sites other than chordoma.(Table 2)

In our patient, primary site was sacrum. Radiographic findings were expansile lytic lesion with soft tissue mass although intratumoral calcification was not present. Histologically a lobular cellular structure with arrangements in cords with ample intercellular mucinous matrix was present. Risk factors for metastasis included younger age group (40 years of age) and the location of tumor (sacral region).

Radiation therapy offers excellent palliation of pain in bone metastases and improves quality of life. Our patient had 90% subjective relief in pain. The overall median survival time with chordoma has been estimated to be approximately 6 years, with a survival rate of 70% at 5 years, falling to 40% at 10 years. Our patient presented with bone, lung and liver metastasis and the metastasis developed after five years of diagnosis. Metastatic disease is considered of adverse prognostic significance because most of the patients die within 40 months.

REFERENCES

1. Papagelopoulos PJ, Mavrogenis AF, Galanis EC, Savvidou OD, Boscainos PJ, Katonis PG, et al. Chordoma of the spine :clinicopathological features, diagnosis and treatment .Orthopedics 2004;27:1256-1263.
2. Gerber S, Ollivier L, Vanel D, Missenard G, Brisse H, de Pinieux G, et al . Imaging of sacral tumours.Skeletal Radiol 2008; 37:277-289.
3. Peh WC, Koh WL, Kwek JW, Htoo MM, Tan PH. Imaging of painful solitary lesions of sacrum.Australas literature 2007;51:507-515.
4. Dahlin DC, MacCarty CS.Chordoma:A Study of fifty nine cases.Cancer 1952;5:1170-1178.
5. Higinbotham NL, Phillips RF, Farr HW, Hustu HO. Chordoma:Thirty-five-year study at memorial hospital.Cancer 1967;20:1841-1850.
6. Fagundes MA, Hug EB, Liebsch NJ,Daly W, Efird J, Munzenrider JE . Radiation therapy for chordomas of the base of skull and cervical spine: patterns of failure and outcome after relapse. Int J Radiat Oncol Biol Phys 1995; 33:579-584.
7. Sundaresan N, Galicich JH, Chu FC,Huvos AG. Spinal chordomas. J Neurosurg 1979; 50:312-319.
8. Chambers PW, Schwinn CP. Chordoma: a clinicopathologic study of metastases. Am J Clin Pathol 1979; 72: 765-776.
9. Keim LW, Hoffman PF. Bilateral symmetrical metastases to clavicular heads from bronchogenic carcinoma simulating primary hyperparathyroidism. Chest 1974; 66:99- 102.