PLASMOCYTOMA OF THE CLAVICLE

Report of a Case

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Solitary plasma cell myeloma of the bone is a rare disease, approximately 50 cases so designated having been recorded in the literature. Only 1 instance of myeloma beginning as an apparently solitary lesion in the clavicle is on record[1] so far as can be determined. In the case to be presented the lesion appeared to be limited to the clavicle except for possible invasion of the sternoclavicular joint. Considering the difficulty of positively ruling out other osseous involvement, the term plasmocytoma, without further qualification, is considered a more accurate designation than solitary plasma cell myeloma.

The following case presents an interesting example of this type of primary neoplasm of the bone.

Case Report

A colored housewife, aged 46, entered Cleveland Clinic on August 2, 1946, to consult Dr. T. E. Jones because of pain and swelling of the right shoulder. Three years before entry she had first experienced pain in the right shoulder with radiation up the neck and behind the right ear, and at that time a diagnosis of arthritis had been made. There was transient swelling over the clavicle. After a year the pain became so severe that she had to limit her activities for two weeks. Following this rest, the pain and swelling subsided. During the two years before entry pain had recurred intermittently, and during the year before admission the swelling had gradually increased in size. Following hard work it enlarged temporarily. At the time of entry the patient stated that she had no pain unless she moved her shoulder or lay down without a pillow.

Except for occasional pain in the joints she had always been well. There had been no weight loss.

Physical examination revealed an obese colored woman, 5 feet 7 inches in height and weighing 248 pounds. Over the right clavicle was a large mass 12 x 9 x 6 cm., firm, fairly smooth, fixed to the medial aspect of the right clavicle and extending to the level of the third rib inferiorly, to the midline medially, and as high as the suprasternal notch. There was no apparent attachment to the thyroid gland. The skin was freely movable over the mass. On movement of the right shoulder the patient experienced slight pain in the region of the mass. Except for extreme obesity, the physical examination was negative. The temperature, pulse rate, and respirations were normal.

Roentgen Examination

The proximal two-thirds of the right clavicle showed extensive bone destruction with some new bone formation and an associated soft tissue mass. The new bone appeared as irregular strands infiltrating into the soft tissue mass. Other bones of the thorax were not involved.

X-ray examination of the chest was not entirely satisfactory. There was no evidence of pulmonary metastasis. The hilar lymph nodes did not appear enlarged. Progress studies were advised.

The roentgenologic diagnosis was malignant neoplasm of the right clavicle, possibly secondary degeneration of a benign tumor, appearance consistent with chondrosarcoma. The tumor mass was regarded as of probable primary origin in the clavicle rather than metastatic, though the latter possibility could not be excluded (fig. 1).

Sixteen days after the first Clinic visit the patient was admitted to the hospital for operation by Dr. T. E. Jones. At this time routine urinalysis was negative. Examination of the blood revealed a red cell count of 3,720,000 with 11 Gm. per cent hemoglobin and a white cell count of 4150. Blood Wassermann and Kahn tests were negative.

Findings at Operation

On the day following admission an operation was performed. An incision was made over the right clavicle from a point over the right shoulder to a point over the attachment of the sternomastoid muscle. The skin and subcutaneous tissues were reflected by sharp dissection, exposing a fusiform tumor of the right clavicle 16 x 8 x 8 cm. The lateral portion of the clavicle was exposed and divided approximately 5 cm. from the acromial

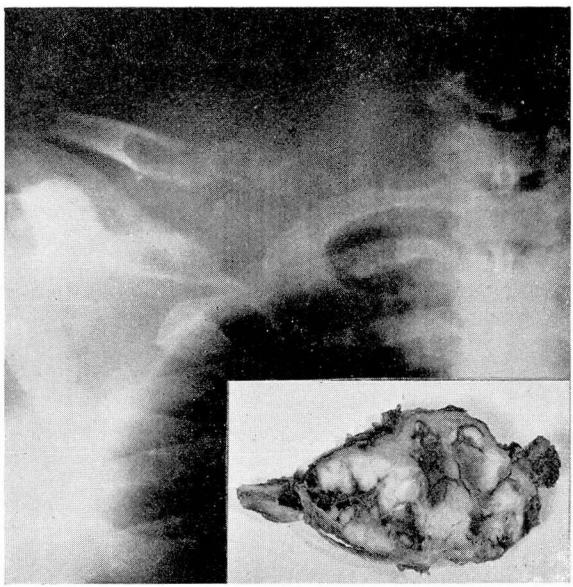

FIG. 1. Roentgenogram of right clavicular area.
Inset. Gross specimen showing replacement of greater portion of clavicle by tumor.

joint. The clavicle was then disarticulated from the sternum. At this point there appeared to be some invasion across the joint, therefore the joint cartilage was excised and a portion of the adjoining sternum curetted away. The area was thoroughly coagulated with the electric cautery. The remaining distal portion of the clavicle was removed by disarticulation at the acromioclavicular joint.

Pathologic Report

Gross examination. The major portion of the specimen was roughly ovoid, measured 12 x 6 x 6 cm., and weighed 250 Gm. The surface was covered by masses of fat tissue and strips of reddish-brown muscle. The tissue of the tumor mass was soft and fleshy in character. The tissue was sectioned with ease but contained irregular bone fragments in several areas. On longitudinal section the bulk of the specimen was formed by a mass 9 x 5 x 5 cm. comprised of pale, "fish-flesh" tissue, friable and soft, with irregular areas, dark reddish-brown in color, and of very soft consistency. There was a marginal zone of fibrous and fat tissue and muscle several millimeters to a centimeter in thickness. A layer of tough, firm, white tissue resembling cartilage or fibrocartilage 2 to 3 mm. in thickness was present medially. A short fragment of bone was present on the lateral aspect. A separate fragment of bone 2.5 x 2 x 1.5 cm. was received separately and appeared to articulate with the lateral aspect of the main mass. Except for a purplish-red, soft zone, 0.5 cm. on its medial aspect, this bone fragment was of hard consistency and white color (fig. 1).

Microscopic examination. Sections from all portions of the tumor mass were of marked cellularity and comprised chiefly of cells of small size, rounded, pear-shaped, or polyhedral, and often with eccentrically placed nuclei. The cytoplasm was basophilic, staining shades of lavender to deep purple in hematoxylin and eosin preparations with a small, oval, pale-staining zone adjoining the nucleus in many cells. Nuclei were round to oval, usually of small size, with very coarse but evenly distributed chromatin. Occasionally a single small nucleolus was present. A few cells contained two or more nuclei, each of similar appearance and size and often with a common clear paranuclear zone. Many cells were within the usual limits of size for normal plasma cells, but some were nearly double that size. Nuclei were often of uniform measurement regardless of cell size but in a few instances were almost double the usual diameter of plasma cell nuclei. Rare mitoses were present; no multiple mitoses or tumor giant cells were found. A moderate number of lymphocytes were scattered among the cells described. There was no intercellular matrix. Frequent capillaries and thin-walled vessels were present throughout the tumor, and cells lay in close relation to the endothelium. Occasionally tumor cells were found in these vessels. Patches of hemorrhage and masses of fibrin deposit were present in portions of the tumor. The external configuration of the tumor tissue was irregular as it adjoined dense connective tissue. However, cells did not stream individually into either connective tissue or muscle and were separated from the latter by a zone of fibrous tissue or periosteum. In a rare section this marginal zone of the neoplasm contained small, homogeneous, pink-staining, hyaline masses which stained positively for amyloid with the Mayer's crystal violet method and were adjoined by occasional giant cells of foreign body type. A rare viable bone fragment was present in the connective tissue about the tumor, and occasionally there was slight osteoblastic activity. In sections taken from the sternoclavicular joint aspect the cartilage was preserved. A few foci of lymphocytes and rare typical plasma cells were found in the connective tissue and muscle.

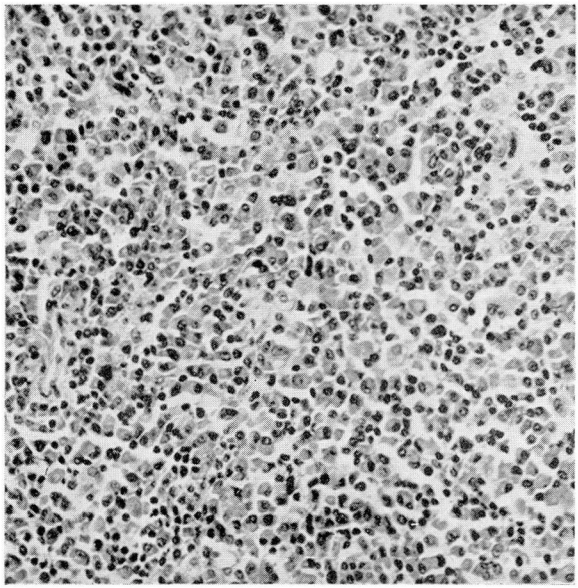

FIG. 2. Photomicrograph showing neoplastic cells with eccentric nuclei and pale staining paranuclear zones. x200.

The pathologic diagnosis was plasmocytoma involving the clavicle and adjoining soft tissue structures (fig. 2).

Postoperative Course

Except for some elevation of the pulse rate and temperature, which returned to normal with penicillin therapy, the postoperative course was uneventful.

Examination of the blood on the ninth postoperative day revealed a red cell count of 3,630,000, hemoglobin 9.0 Gm. per cent, and a white cell count of 9700 with 65 per cent neutrophils, 24 per cent lymphocytes, 8 per cent monocytes, 1 per cent eosinophils, 1 per cent basophils, and 1 per cent metamyelocytes. No abnormal forms were observed, and the platelets were normal. There were 2.9 per cent reticulocytes. A sternal marrow examination revealed cellular marrow with a relative increase in myeloid elements and with a slight shift to the left in this series. Plasma cells were slightly increased to 3.5 per cent and showed moderate pleomorphism, but the majority of nuclei were of mature type and no extremely large forms were seen. Dr. L. W. Diggs examined the marrow and did not regard it as diagnostic.

Roentgenologic examination fourteen days after operation revealed no evidence of myeloma in plates of the skull, lumbar spine, sternum, right hip, and right knee.

No examination of the urine for Bence Jones protein was made at this time.

The patient was discharged on the fifteenth postoperative day and advised to return every three months for check-up.

A roentgenogram of the right clavicle two and one-half months after discharge revealed no evidence of a soft tissue mass in the original site of the clavicle.

Six months after discharge the patient was free of symptoms. Hemoglobin was 12.5 Gm. per 100 cc., the white cell count 8000 with 59 per cent neutrophils.

Nine months after operation a check-up revealed no radiologic evidence of local recurrence in the right clavicular area and no apparent myeloma of the skull, lumbar spine, sternum, or pelvis. X-ray examination of the chest was negative except for absence of the right clavicle. The findings in a sternal marrow examination were essentially as before (2 per cent plasma cells). No Bence Jones protein was demonstrable in the urine. The serum total protein was 6.9 per cent. A sedimentation rate was somewhat increased to 1.0 mm. per minute (normal limit 0.45 mm. per minute). The patient was free of symptoms.

Comments

Plasmocytoma occurring as an apparently solitary lesion without detectable involvement of other bones has been described in many anatomic locations. In 1940 Paul and Pohle[2] reviewed 40 cases from the literature and added 5 of their own. They found the most common locations to be the bones of the pelvis (10 cases), dorsal spine (9 cases), femur (8 cases), and humerus (5 cases), with other recorded locations in the jaw, tibia, cervical region of the spine, skull, and clavicle. The average age of patients was 48 years; two-thirds were of male sex. Unostotic location of the tumor in some of the cases can be only presumptive, as in several instances foci in other bones were recorded only a few months after the original lesion was discovered. Cutler *et al.*[3] divide the solitary plasma cell myelomas into two groups, one in which the primary lesion is followed in several months or several years by typical multiple myeloma and a second group in which the lesion apparently remains solitary. Unfortunately the time of follow-up has been too short to evaluate adequately the course of many of the recorded cases. However, the above authors[3] report 3 patients followed for periods of four, nine, and ten years, respectively, without appearance of tumor in bones other than the primary site. Pasternack and Waugh[4] report a case of solitary myeloma of the humerus under observation seven and one-half years without evidence of generalized bone involvement. Bailey[5] presents a case followed through seven years without diffuse spread. Stewart and Taylor[6] record 2 instances of eight-year survival without evident diffuse skeletal involvement. At the time of first observation there is no means of determining which course these tumors will follow. Diffuse involvement may occur several months or even several years after discovery of the solitary lesion. Jacox and Kahn[7] record a case in which the interval was four and one-half years.

The first manifestation of the lesion may be pain, presence of a mass, or spontaneous fracture. Pasternack and Waugh[4] in a review of 30 cases found pain the presenting symptom in 74 per cent, swelling in 48 per

cent. Tennent[8] found a high percentage of pathologic fractures when the primary lesion occurred in the long bones.

Positive diagnosis of this tumor before histologic examination is difficult. The radiologic appearance is variable, as the lesion may simulate giant cell tumor, bone cyst, metastatic carcinoma, chondroma or chondrosarcoma, and hemangioma of the bone. Generally the diagnosis of giant cell tumor or metastatic carcinoma is made roentgenologically. The excellent review by Paul and Pohle[2] includes detailed discussion of radiologic features. Bence Jones proteinuria usually is absent until there is evidence of polyostotic involvement. Blood protein levels are usually normal. The sedimentation rate is not increased usually until there is diffuse disease. Sternal marrow examination is said to be most valuable. In the presence of generalized involvement there will usually be abnormalities in the percentage and, perhaps, the type of plasma cell, and one report states that the sternal marrow examination may be the only examination of value when the lesion appears to be solitary.[8] Bichel and Kirketerp[9] report an increase in plasma cells of the sternal marrow to 42.2 per cent and 22 per cent, respectively, in 2 cases in which radiologically there were solitary lesions. In these instances there were blood protein changes consistent with multiple myeloma, and in 1 instance Bence Jones protein was also found. They call attention to the fact that diffuse disease may be present even when there appears to be only a single lesion on radiologic examination. In the instance of the present report the plasma cells of the sternal marrow were slightly increased to 3.5 per cent, but this was not regarded as diagnostic. The prognostic importance of such a level cannot yet be judged.

There is an insufficient number of cases reported with adequate follow-up to evaluate therapy properly. The natural history of the disease is such that generalization may not be evident for several years. In a case reported by Tennent[8] in which no treatment was given, two years elapsed before diffuse involvement was observed. Any judgment of therapeutic effectiveness must take this into consideration. Excision or amputation, curettage, radiation therapy, or radiation therapy in addition to one of the above have all at times been associated with survivals of two years or more. For solitary lesions of the extremities Paul and Pohle recommend radiation and state that excision or amputation should be considered in the event of recurrence. For other locations they advocate curettage and radiation, where feasible, or radiation alone. For recurrent lesions after surgical intervention, radiation therapy has shown a palliative effect and perhaps has prolonged life. When used as primary therapy it has relieved pain or other discomfort. The lesion decreases in size but generally does not disappear. In the tabulation of

45 cases by Paul and Pohle there are 9 instances of survival for more than four years. Three of the patients received radiation therapy following curettage or partial resection, 1 was given roentgen therapy after biopsy, and 5 were treated by resection or amputation. In the last group, surgical intervention occurred after radiation in 2 instances. In the case here reported, local surgical excision of the mass was elected and reliance placed on such removal. There has been insufficient time for determination of the result.

Summary

Plasmocytoma of the bone manifested as a solitary lesion is rare but has been established as an entity. Though in many instances the disease has terminated with diffuse involvement of the skeleton, it has been recorded as remaining localized or not recurring for a period of years. Sternal marrow studies may be of aid in the diagnosis or in determining generalization. The lesion is first manifested by pain, swelling, or, as is common in the long bones, spontaneous fracture. Radiologically the solitary lesion most often simulates giant cell tumor or metastatic carcinoma. Resection, when feasible, seems the most logical therapeutic approach. From published reports radiation therapy appears to be of value in the control of recurring lesions and when used primarily may reduce the size of the lesion and alleviate symptoms.

A case of apparently solitary plasmocytoma of the clavicle is presented.

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