INTRODUCTION

Chordoma, or more properly chordocarcinoma or chordoepithelioma, although a rare tumor, is becoming better known. Up to the present time upwards of 100 cases have been recorded in the literature, and the writers are familiar with a number of others through personal communications.

Luschka in 1856, Virchow in 1857, and Müller in 1858 were the first to describe chordoma dorsalis under the caption "ecchordosis physaliphora." It was Müller who first considered it to be of notochordal origin, a view fully confirmed by Ribbert in 1894. The latter reported that 2 per cent of all cases autopsied by him had notochordal remains in the region of the clivus. Occasional cases were recorded up to 1922, when Stewart reviewed the literature and collected 26 cases; in 1924 Coenen collected 68 cases. Since that time case reports have become more common, indicating increased knowledge and interest in this subject.

Some pathologists have remained up to the present somewhat skeptical of the existence of chordoma as a definite entity. Ewing in "Neoplastic Diseases" (3d. Ed., page 212) makes the following statement: "The positive identification of chordoma is not readily accomplished. Two common tumors closely simulate, or even duplicate the structure attributed to chordoma, viz., myxochondroma and colloid carcinoma of the intestinal canal. . . . I have seen these features, except the broad attachment to the sacrum, in colloid carcinoma of the rectum. In the pharyngeal region atypical chondromas are rather common and must be considered a more probable occurrence than chordoma." The late Dr. A. S. Warthin was for a time skeptical about the existence of chordoma as a tumor entity. To quote from a statement by Dr. C. V. Weller, who succeeded Warthin as Professor of Pathology at the University of Michigan: "For a number of years Dr. Warthin took a neutral stand on the existence of chordoma as a tumor
entity. For a number of years before his death, however, he accepted this neoplasm as such and made the diagnosis himself on a number of occasions. It was particularly in the period 1920–1925 that he used to question whether chordoma should be separated from chondroma.”

By far the majority of cases have been reported as from the spheno-occipital region or the sacrococcygeal region. In the former region the origin is from the clivus blumenbachii in most instances. Of the cases reviewed by Stewart (13), 15 were from the spheno-occipital and 9 from the sacrococcygeal region. Coenen (4) found 36 cases from the spheno-occipital and 25 from the sacrococcygeal region. A few cases have been recorded as having arisen from heterotopic foci, as the upper and lower jaws, left superior occipital region, and nasopharynx.

Cases arising from the cervical, dorsal, or lumbar spine form a small part of the recorded group. The purpose of this paper is to report a case originating from the cervical region of the spinal column and to review the cases in the literature arising in this same area.

CASE REPORT

P. G., an Italian male, a cement worker, aged fifty-two, entered Grace Hospital Oct. 16, 1931, complaining of pain in the neck, both shoulders, and both arms, more pronounced on the left side.

Present Illness: The onset had been gradual, dating back about eighteen months, and the condition had become progressively more severe. The pains were paroxysmal, sharp and lightning like, shooting down the shoulders and outside of the arms. Of late there had been much loss of sleep. There was also progressive weakening of the upper extremities, particularly of the left. Occasionally there were severe cramps in the legs on walking.

General Physical Examination: The subject was a well developed, muscular man, holding his neck rigid. No masses were palpable in the neck. There was some tenderness in the midportion of the cervical spine. The heart and lung findings were negative; the blood pressure was 130/30. The abdominal findings were also essentially negative. Rectal examination revealed no abnormalities; the prostate was small, soft, and not tender.

Muscular System: There was atrophy of the deltoid, supraspinatus, and infraspinatus muscles on both sides, more marked on the left. There was some atrophy of the left hypothenar eminence, but no atrophy or deformity of the lower extremities. There was definite weakness of the grip on the left side. Some impairment of coordination of the movements of the fingers was observed, more on the left than on the right.

Reflexes: The pupillary reactions were normal. The left biceps reflex was absent, the right diminished. The triceps reflexes were present on
both sides. The abdominal reflexes were present in three quadrants and absent in the left upper quadrant. Cremasteric reflexes were present. The knee and achilles jerks were hyperactive, especially on the left.

Sensations: There was diminution of heat, cold, and pain sensation below the 5th cervical dermatome. The vibratory sense was absent over the left clavicle and diminished in the left elbow, knee, and foot. On the right side it was diminished at the elbow, almost absent in the hand, and diminished in the leg but to a lesser degree than on the left. Sense of position and passive movement was lost in the fingers of both hands and diminished in the legs, particularly on the left.

Gait: There was no Romberg sign. No evidences of spinal or cerebellar ataxia were observed. The finger-to-nose test and heel-to-knee test were normal. On walking, the patient held the neck rigid and slightly tilted to the left.

Laboratory Findings: The spinal fluid manometric readings were suggestive of blocking of the cerebrospinal fluid circulation. The fluid was entirely normal. The blood contained 105 per cent hemoglobin (Sahli), 5.25 million red cells, 8.8 thousand white cells per cu. mm.,

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**Fig. 1. Lateral View of Cervical Spine before Operation**

Note the destructive lesion in the body of the fifth cervical vertebra.
FIG. 2. BIOPSY SPECIMEN: LOW-POWER VIEW

FIG. 3. BIOPSY SPECIMEN: HIGH-POWER VIEW

Note lack of stroma, phyaliphorous cells, and marked variation in size and staining of nuclei.
polymorphonuclear neutrophils 71 per cent, lymphocytes 24 per cent, large mononuclears 5 per cent. The blood sugar (dextrose) was 105 mgm., the non-protein nitrogen 34, urea nitrogen 15, creatinine 1.5 mgm. per 100 c.c. The stomach contained 30° free HCl and 52° total acid following an Ewald test meal; it contained no blood, bile, or lactic acid.

X-Ray examination of the cervical spine (Fig. 1) showed a destructive process in the body of the fifth cervical vertebra. It simulated a metastatic new growth in appearance, although cervical tuberculosis was considered.

**Fig. 4. Lateral View of Cervical Spine Five Weeks after Operation, Showing Extension of Process**

Operation (E.S.G.): In view of the severe pains in the limbs and the presence of spinal fluid block, it was thought justifiable to do a decompressive laminectomy and also a section of the sensory roots in the areas of distribution of the pain. Operation was done on Nov. 4, 1931. The spinous processes of the 4th, 5th, and 6th cervical vertebrae were removed. The normal dural pulsations and the peridural fat were absent. The dura was incised, the cord was exposed and examined. There was no demonstrable macroscopic change in the nervous tissue elements. The body of the fifth cervical vertebra was soft and spongy and bulged
slightly posteriorly. A small fragment was removed for tissue study. There was no free fluid or pus. The lateral processes of the vertebrae being solid, fusion was not deemed necessary. The fifth and sixth sensory nerve roots on the left were cut to relieve the pain.

Microscopic Examination of the Tissue (Figs. 2 and 3): The fragment of tissue was fixed in formol, embedded in paraffin, and stained with hematoxylin and eosin. The amount was so small that only a few sections could be made, and no special staining could be done. The specimen contained no fibrous tissue stroma. It was composed of cells varying markedly in size. The nuclei varied in their staining qualities, some staining deeply, others faintly; a few cells contained more than one nucleus. The protoplasm of nearly all of the cells was physaliphorous, and contained many vacuoles of varying sizes, some pressing upon and distorting the nuclei. In addition there appeared considerable intercellular vacuolization. A diagnosis of chordoma dorsalis was made.

Postoperative Course: Following the operation the patient was completely immobilized for two weeks, with slight traction. Afterward the neck was immobilized by means of a head and body spica. For about a month after the operation he was quite comfortable. He then began to complain of severe pains, as before the operation. On Dec. 7 a second roentgenogram was taken (Fig. 4), which showed a definite extension of the process, with the 6th cervical vertebra undergoing destruction. Deep x-ray therapy was given with the hope of alleviating the condition. This apparently had no effect. The pains continued as before, and the patient lost weight. At the present writing (June, 1932), he is suffering severe pain and refuses to have further nerve sectioning because of the risk of operation.

Review of Literature

Careful search of the literature revealed eight previously reported cases which we believe may be classed as chordomata arising from the cervical region of the spine. Some which have been considered as arising in this region we feel should not be included because of insufficient proof; these are mentioned later. Undoubtedly a number of the nasopharyngeal cases have arisen from the cervical region of the spine, but no attempt has been made to classify these. The cases are given chronologically.

Case 1: This case was recorded by Trélat (15) in 1868 and was referred to by Desjacques (5) in 1927, in his article on chondromas of the spine. It was reported as a chondroma, but Desjacques believed it to be a chordoma, and we accept his opinion. The patient was a woman, aged twenty-seven, who had had severe pains in the right arm for eighteen months, and a hard mass occupying the lateral aspect of the neck for a year. Examination showed the mass to be under the sternomastoid muscle on the right, extending from the angle of the jaw down to the clavicle. The overlying skin was movable. The carotid sheath and
The right radial pulse was weak and there were dilated veins in the right arm. With a trocar a small piece was removed, which Ranvier called "enchondrom muquez." Later a second operation was performed and the mass was found to be adherent to the spinal column in the cervical region, where, apparently, it had originated. There is little doubt that the tumor was a chordoma arising from the cervical spine, although we do not know the exact point of origin.

Case 2: Klebs (10) described a case in 1889 which he examined for Heine of Prague. The tumor arose from an intervertebral disc in the cervical region of the spine. Klebs called it a chondroma and stated that it contained cells which were identical with the cells found in tumors arising from the clivus and with those found in the embryonic notochord. His diagnosis was "chordal chondroma."

Case 3: Steiner (12) of Cologne reported a case in a boy of sixteen, a student. He complained of pain in the back of the neck and soreness on pressure in the upper cervical region. Six months later paralysis of the left arm developed with claw-like position of the fingers; also paralysis of the left foot, with foot clonus. A clinical diagnosis of tuberculous spondylitis was made and an extension dressing was applied. The symptoms progressed, however. The left eye and the tongue deviated to the left; bilateral choked disc and dysphagia developed. Death occurred suddenly while the patient was entirely conscious.

Autopsy showed a calcareous tumor which extended from the height of the clivus through the occipital groove to the axis. It compressed the medulla and the spinal cord to the size of a lead pencil. The abducens nerve and apparently the hypoglossal were surrounded by the growth. The tumor was most intimately connected with the odontoid process of the axis, and Steiner considers this as its origin. Fischer (7), who examined the tumor, was inclined to doubt that it arose from the axis, but Coenen in his review accepts it as having arisen from the odontoid process of the axis, and classes it as "dentate" in location. Fischer reported it to be a chordosarcoma.

Case 4: This case was described by Syme and Cappell (14) in 1926. The subject was a male, aged fifty-nine, who complained of pain, of two months' duration, on moving the head from one side to the other and in the anterior posterior direction. There were lateral stiffness, some difficulty in swallowing, and some interference in breathing. Speech, also, was affected. A swelling was present in the throat, on the posterior wall of the pharynx, extending into the nasopharynx and down into the laryngopharynx. It was more prominent on the left side, and the left posterior pillar of the fauces was stretched over it, but there was no ulceration of the mucosa. It was firm on pressure, somewhat lobulated, and negative on aspiration. On operation the tumor was found to be attached to the vertebral column anteriorly. It invaded the 2d, 3d and 4th cervical vertebrae. Microscopic examination revealed a typical chordoma. At a second operation, six months later, the growth was less well defined, and reached from the base of the skull down to the fifth cervical vertebra.
CASE 5: This case was recorded by Cappell (2) in 1928. The subject was a male, aged forty, who complained of pain in the neck and weakness of the arms of six months' duration. Examination on admission, four weeks before death, revealed a well-marked wasting of the muscles of the shoulder girdle, especially of the trapezius and deltoids. There was marked loss of power in the arms, and the grip was weak, but the intrinsic muscles of the hand were not wasted. The biceps jerk was diminished, but the other arm reflexes were increased; the knee jerks and ankle jerks were increased. Patellar and ankle clonus were present on both sides, with extensor plantar response. Five days before death sudden loss of power in both arms appeared, together with complete loss of sensation for pain and temperature in the trunk, arms, and legs. Paralysis of the bladder and rectum followed, with incontinence.

At autopsy an ecchordosis physaliphora 3 mm. in diameter was found at the clivus. The spinal cord was found to be flattened and compressed at the level of the 3rd, 4th, and 5th cervical vertebrae by a tumor mass which was adherent to the anterior and lateral aspects of the dura mater. It was found that the growth had originated in the body of the 4th cervical vertebra, and had extended to the 3rd, 5th and 6th cervical vertebrae. Microscopic examination showed it to be a typical chordoma.

CASE 6: This case was recorded by Andrée-Thomas and Villandre (1) in 1928. The subject was a female, aged fifty-eight, who complained of pains in the neck, right hand, and mid-cervical region. Roentgenograms showed changes in the 4th and 5th cervical vertebral bodies which were thought to be of a tuberculous nature. The patient was immobilized but did not improve. Eight months later there was enlargement of the neck. Complete paralysis of the left leg, partial paralysis of the right leg, and almost complete paralysis of the arms developed. There was slight involvement of the sphincters. Operation was performed, and microscopic examination of the material removed showed a typical notochordoma.

CASE 7: This case was recorded by Chiari (3) in 1928. The subject was a female thirty-five years of age who had had pain in her right arm, neck, and shoulders for eighteen months. She was admitted with total paralysis of the left arm and partial paralysis of the right arm. The reflexes of the lower extremities were markedly exaggerated. Loss of motion of the lower extremities, with hypesthesia and almost complete hypalgesia from the 4th cervical vertebra downward, developed. Death occurred three weeks later from sepsis due to a deep sacral abscess.

At autopsy the spinal cord in the region of the 4th to 6th cervical vertebrae was found to be compressed by an extradural tumor. The tumor was connected with the 5th cervical vertebra by a pedicle. At the clivus there was a typical chordoma pearl the size of a pea. It appeared that the tumor arose from the body of the fifth cervical vertebra, and not from an intervertebral disc. The microscopic picture was that of a chordoma.

CASE 8: Herrmann (9), in 1929, reported a case in a man of forty-four who complained of pain in the neck and occiput, inability to turn the head without pain, and a cracking sensation on motion of the head
and neck. He was unable to stoop over and his head felt as if it were too heavy. The neck was stiff, and there was a tendency to bend the head forward and to the left. The posterior pharyngeal wall was arched forward, as occurs with retropharyngeal abscess. The swelling appeared to arise from the upper two cervical vertebrae. It was firm and elastic, and no fluctuation could be demonstrated. A wedge-shaped piece of tissue was removed through the mouth, and the microscopic sections showed chordoma. X-ray pictures suggested tuberculosis, with destruction of the axis and atlas.

There are two other cases worthy of mention, since it may be that they represent chordomas of cervical vertebra origin. The first is that of Linck (11). This patient presented symptoms of middle ear suppuration together with retropharyngeal swelling. The ear was operated upon, and the retropharyngeal swelling punctured without result. Later, at a second operation, slimy material was removed which proved to be a malignant chordoma when sectioned. There was no evidence as to its exact origin. The second case is that of Fabricius-Moller (6). This patient had nasal obstruction and thickness of speech, and a swelling was found filling the posterior pharyngeal wall and extending below the level of the soft palate. An incomplete operation was performed, and the growth recurred in a few months. The patient remained fairly well, however, for four and a half years, when he again returned to the hospital with a large tumor in the posterior pharyngeal region. A second operation was performed, and the tumor was removed as far as possible. Six months later the patient was still well, although some swelling was present in the posterior pharyngeal wall. This tumor, also, has been described as of cervical origin, although in the original article no such claim is made.

**COMMENT**

Chordoma is a more frequent condition than is ordinarily supposed. It has been well proved that its origin is from embryonic remains of the primitive notochord. In the adult the normal structure persisting from the notochord is the nucleus pulposus, found in the intervertebral discs. Chordomata, however, arise from other notochordal foci as well, both in the spinal column, at the cephalic and caudal extremities, and from heterotopic foci. Those which arise from the cervical spine are rare, there being reports of but nine cases, including the case reported herein, in the literature. In two cases (Cases 3 and 8) the odontoid process of the axis was the site of origin. In one case (Case 2) the origin was from an intervertebral disc (nucleus pulposus?); in four cases from one of the vertebral bodies (Case 4 from the 3d; Case 5 from the 4th; Case 7 and the case here reported from the 5th). In two cases, the exact origin was unknown (Cases 1 and 6). In all the
cases that were examined by x-ray the diagnosis of tuberculosis was made, although in our case secondary cancer was suspected. We believe, from this standpoint alone, that in the consideration of tuberculosis of the cervical spine, chordoma must always be kept in mind as a possibility. There seems to be no typical x-ray picture of chordoma in the cervical region.

In Cases 1, 4, and 8 the tumor grew anteriorly, so that a tumor mass was present. In Case 1 the tumor was in the lateral aspect of the neck, and in cases 4 and 8 there was a tumor mass projecting into the pharynx. In Case 3, and in our own case, there was mention of tenderness in the cervical region of the spine. In Cases 3, 5, 6, 7, 8, and in our own case there were definite symptoms of spinal cord tumor as manifested by spontaneous pains due to nerve root infiltration, and paralysis and atrophy of groups of muscles. There was proved compression of the spinal cord in Cases 3, 5, and 7, demonstrated by post-mortem examination.

We tried deep x-ray therapy, but in the light of the recent monograph by Grinda (8), in which he reports sacrococcygeal cases treated by deep radiation, we should expect little in the way of result. After three months, our case seems to substantiate this belief.

It would seem that in cases of tuberculosis of the spine in which operation is performed, a biopsy should be done. In our case the diagnosis would have been missed except for biopsy.

**Summary**

A case of chordoma of the body of the fifth cervical vertebra is reported, and a review of reported cases believed to have originated in the cervical region of the spine is presented.

**Bibliography**

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