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Histological Study on Local Invasiveness of Clival Chordoma. Case Report of Autopsy

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Summary

Background. It is well known that clival chordomas invade bony structures; however, their invasiveness of other structures has rarely been reported. We report an autopsy case of clival chordoma in a 61-year-old woman, who underwent surgery 6 times for a period of 8 years, with special reference to its local invasiveness.

Method and Findings. The histological studies showed that the tumour grew in the loose connective tissue in multilayers or multilobular fashion, invading the submucous layer in addition to the bone; however, it did not invade other soft tissue, particularly vital neurovascular structures or the dura itself, even in the advanced stage.

Interpretation. The dura may serve as a strong barrier against tumour invasion into the intradural space, which emphasises the importance of not injuring the dura, when the tumour is removed extradurally. For radical removal of clival chordoma, it will be necessary to remove the tumour extensively including normal bone and soft tissues surrounding the tumour, especially the mucous membrane, because the normal mucous membrane is surgically difficult to preserve by dissecting it from the invaded submucous layer.

Keywords: Chordoma; clivus; tumour invasion; autopsy.

Introduction

It is well known that clival chordomas invade the bony structure [25]; however, their invasiveness other structures has rarely been reported [7, 10, 12, 21]. We report an autopsy case of clival chordoma with special reference to its local invasiveness.

Methods and Patient

Case Report

Clinical course. This 61-year-old woman was referred to our hospital at the age of 53 years, with a 1.5-year history of diplopia and left-sided ptosis, and a 3-month history of numbness of the face on the left side. Neurological examination on admission revealed bilateral visual disturbance, left-sided ptosis and total ophthalmoplegia, and hypalgesia and hypesthesia of the face in the left side. Computed tomography scanning revealed a low density mass with enhanced rim at the left petroclival region as well as bony destruction of the upper clivus, petrous apex and anterior clinoid process (Fig. 1, left).

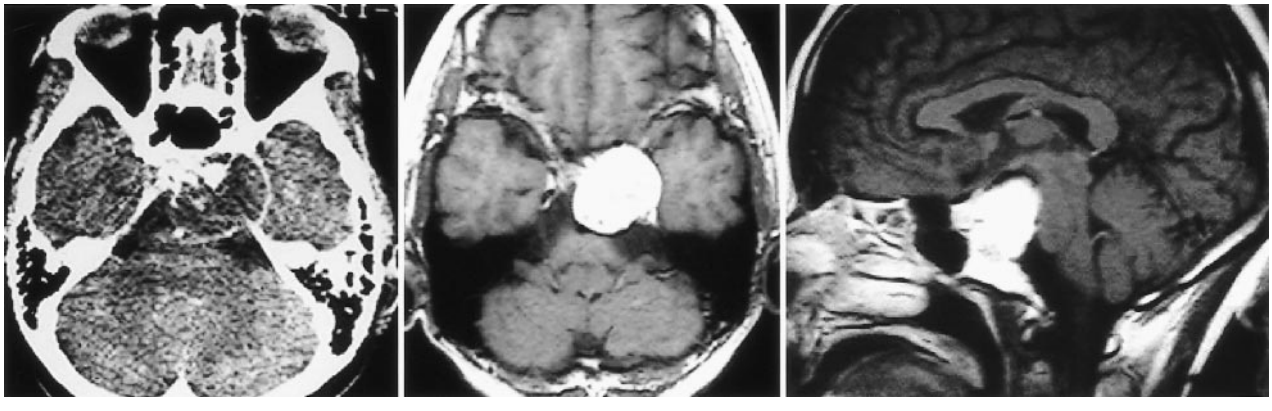

Fig. 1. Neuro-imaging before the first operation. A computed tomography scan revealing a low density mass with enhanced rim at the left petroclival region as well as bony destruction of the upper clivus, petrous apex and anterior clinoid process (left). Magnetic resonance images showing a well-enhanced mass involving the left cavernous sinus and clivus (middle: axial view, right: sagittal view)

Table 1. *Profile of Surgery in the Patient*

	Date of surgery	Procedures
1.	1989, March	It. frontotemporal craniotomy with zygomatic osteotomy
2.	1990, August	transsphenoidal approach
3.	1991, June	transfacial approach
4.	1992, October	It. infratemporal fossa approach
5.	1995, March	transbasal approach
6.	1997, April	rt. infratemporal fossa approach

It. Left, *rt*. right.

Magnetic resonance (MR) imaging revealed a well-enhanced mass as low signal intensity in T1-weighted image and high signal intensity in T2-weighted image involving the cavernous sinus and clivus (Fig. 1, middle and right). The tumour was subtotally removed via a left frontotemporal craniotomy with zygomatic osteotomy. Left visual acuity improved slightly; facial hypalgesia and hypesthesia were slightly worsened postoperatively. Pathological diagnosis was typical chordoma.

After the operation, the patient underwent surgery 5 more times for tumour regrowth, each time resulting in partial removal (Table 1). In April 1997, the 6th operation was performed via a right infratemporal fossa approach. Pre-operative examination showed bilateral blindness, cranial nerve palsy of the bilateral 3rd to 6th nerves and right 12th nerve, and right-sided slight hemiparesis without sensory disturbance or cerebellar sign. The tumour was partially removed uneventfully. An MR image taken 6 months after the surgery revealed massive regrowth of the tumour at the central external skull base extending to the right infratemporal fossa. It was encasing both internal carotid arteries, and compressing the midbrain (Fig. 2). One month later, she died due to respiratory dysfunction.

At autopsy, the tumour was removed en-block with surrounding structures. This specimen was then studied macro- and microscopi-

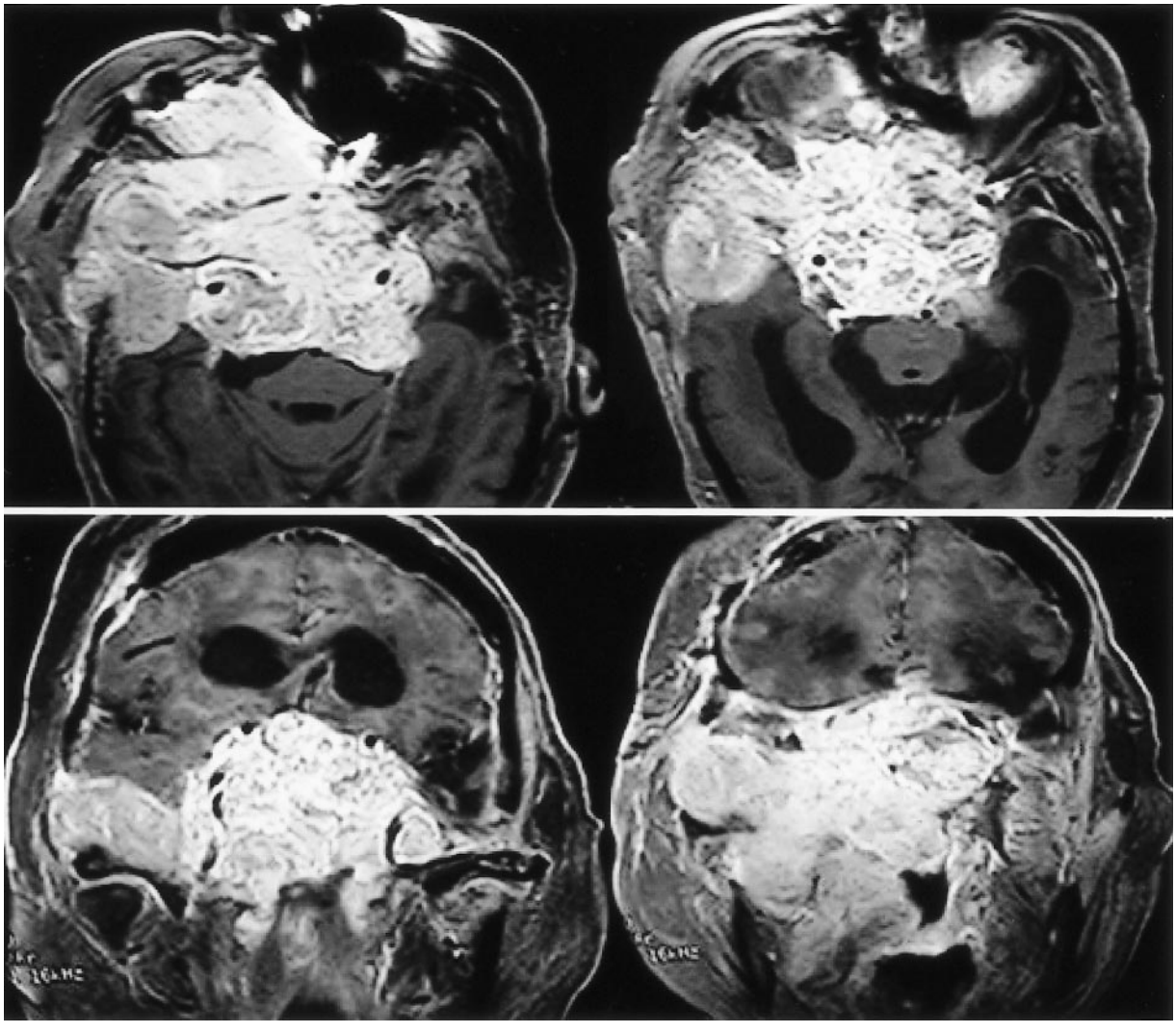

Fig. 2. Magnetic resonance images before the patient's death. Enhanced magnetic resonance images obtained at 6 months after the last operation (one month before death) revealing massive regrowth of the tumour at the external central skull base extending to the right infratemporal fossa, which is entirely encasing both internal carotid arteries and compressing the midbrain. (upper: axial views, lower: coronal views)

cally with reference to tumour invasion of surrounding structures such as the bone, muscle, mucous membrane, artery, vein, nerve and dura.

Results

Macroscopic Observation

The tumour occupied both cavernous sinuses with extension into the clivus area, adhered moderately to the brain, and compressed the brainstem markedly. The tumour was mostly located extradurally; it partly extended into the intradural space through a ruptured part of the dura mater.

Microscopic Observation

The pathological diagnosis was typical chordoma (Fig. 3.1), which was confirmed by immunohistochemical studies. The tumour grew into the space of the loose connective tissue among the vessels, nerves and muscles in multilayers or multilobular fashion. The loose connective tissue was lost where the tumour had spread. The bone including the marrow was directly invaded by the tumour; the normal bony structure was destroyed with loss of the periosteum, and normal bony margins were unclear without a definite membranous structure between the tumour and bone (Fig. 3.2). The mucous membrane of the nasopharyngeal cavity, the mucous epithelium and propria were compressed without tumour infiltration, while the submucous layer was invaded by the tumour in multilobular or cluster fashion (Fig. 3.3).

The tumour did not invade the artery (internal carotid artery), vein, nerve, muscle or fascia; the adventitia of the artery and epineurium were well preserved, even though these structures were strongly compressed by the tumour (Figs. 3.4–3.6). In this area the tumour was surrounded by a membranous structure, which was not a true tumour capsule (pseudocapsule [19]). The dura mater was compressed and thinned by the tumour, but the dural tissue itself was well preserved and not infiltrated by the tumour (Fig. 3.7).

Discussion

Chordomas, particularly clival chordomas, have been considered as one of the most difficult tumours to treat surgically [5, 8, 13, 18, 19], because of their critical location and infiltrative propensity into surrounding structures. In this study, we found that the tumour tended to grow in the loose connective tissue among the vessels, nerves and muscles in multilayers or multilobular fashion [5, 13, 19], and to invade the bony structure and submucous layer, but did not invade the artery, vein, nerve, dural tissue, muscle, fascia or mucous membrane itself.

Some authors have described that cranial chordomas simply displace surrounding soft tissues without invasion, though they invade the bone [7, 12, 13, 21], or that they only compress the adjacent artery and nerve without invasion [10]. Our observation histologically confirmed most of their findings, but an especially noteworthy finding in our study was that the tumour invaded the submucous layer, and did not invade the dura itself and vital neurovascular structures even in the advanced stage, or even if the repeated surgical interventions have affected the invasive potential of this tumour. Intradural tumour extension, which is occasionally encountered and can bring a fatal result, might be caused by rupture of the dura as a result of tumour compression, not by direct tumour invasion of the dura. The dura serves as a strong barrier against tumour extension into the intradural space. It is, therefore, important not to tear or open the dura, when the tumour is removed extradurally.

Since many skull base approaches to the clival region have been reported [2, 3, 4, 6, 11, 14–17, 20, 22–24], an extensive and aggressive surgical approach is advocated to increase the rate of radical removal [1, 9]. However, the recurrence rate seems to be still high even using advanced microsurgical and skull base techniques [9]. The difficulties in radical removal of clival chordomas may be mainly due to their invasiveness of the submucous layer and extension into the loose connective tissue in multilayers or multilobular fashion. For radical removal of clival chordomas, it will be necessary, at the initial surgery, to remove not only the invaded bone and submucous layer but also normal bone [21] and soft tissues surrounding the tumour, especially the mucous membrane over the tumour as much as possible. Because normal bony margins are not clearly defined [21], the tumour extends in the loose connective tissue, and the normal mucous membrane is surgically difficult to preserve by dissecting it from the invaded submucous layer.

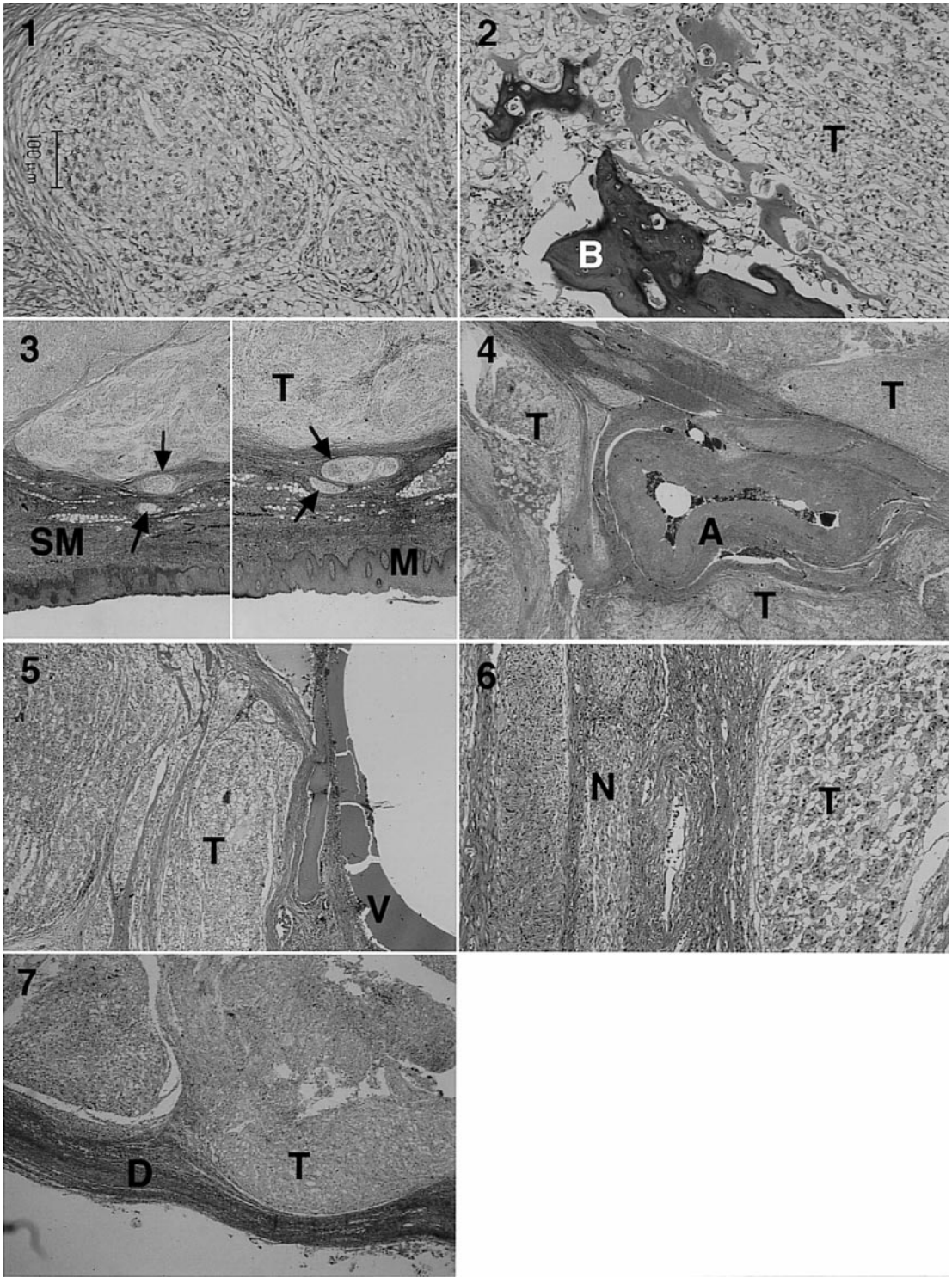

Fig. 3. Microscopic findings of the tumour and its surrounding structures (H & E) (1) showing the pathological findings of the tumour as typical chordoma. Original magnification × 40. (2) showing that the bone is invaded by the tumour (*T*) and its normal structure is destroyed without clear bony margins. Original magnification × 36.7. (3) showing that the epithelium and propria of the nasopharyngeal mucous membrane (*M*) are well preserved but the submucous layer (*SM*) is invaded by the tumour (*T*) in multilobular or cluster fashion (arrows). Original magnification, left × 8, right × 10. (4) showing that the adventitia of the internal carotid artery (*A*) is well preserved without tumour infiltration despite the fact that it is encased by the tumour (*T*). Original magnification × 5.3. (5) showing that the vein (*V*) has no tumour (*T*) infiltration. Original magnification × 13.2. (6) showing that the trigeminal nerve (*N*) has no tumour (*T*) infiltration. Original magnification × 31.2. (7) showing that the dura (*D*) is well preserved without tumour (*T*) infiltration, though it is thinned by tumour compression. Original magnification × 40

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Comment

A well written, interesting Case Report of an autopsy study of a massive chordoma of the base of the skull. The authors concentrate on demonstrating tissues that do not seem to be invaded by tumour. From the imaging it could be quite a unique case.

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