"Tailored" approach to selected recurrent cranio-cervical chordomas: experience and lessons learned*

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Abstract

Background: Among chordoma patients, recurrent cases are by far more complex to be managed, and cranio-cervical junction (CCJ) localizations represent a particular challenge due to the complexity of the anatomical region which makes it difficult to obtain a radical resection.

Methodology: We report our personal experience in treating four patients with recurrent CCJ chordoma with "personalized" multiportal and eventually multistage approaches.

Conclusions: Endoscopic endonasal approaches have gained widespread acceptance and are considered the workhorse in most cases of craniocervical junction chordomas. Nonetheless, in some cases of recurrence, or in presence of very lateralized lesions/ anatomical variations midline approaches are either contraindicated or very difficult to perform. In all these cases it seems reasonable to consider a versatile strategy including different approaches, modulating the surgical needs with different answers and solutions offered by the different routes. In other words to personalize as much as possible the approach, being creative and not dogmatic.

Key words: chordoma, craniocervical junction, recurrent, multistage approach, multiportal approach

Introduction

Chordomas are rare bony lesions arising from notochordal remnants. From a histological point of view they are considered as benign tumours, but their biological behaviour is far from being benign given their tendency to recur and even metastasize. As it is well known, the natural history of the chordoma disease is characterized by multiple recurrences and tumour-related factors are generally the cause of death [(1)]. To current knowledge the best treatment is the association between maximal safe resection and adjuvant radiotherapy [(2)]. Among chordoma patients, recurrent cases are by far more complex to be managed, and cranio-cervical localizations represent a particular challenge. This is due to the complexity of the anatomical region which makes it difficult to obtain a radical resection. Furthermore, it should also be noted that even in cases where it is believed that a "radical" resection has been obtained, it is not uncommon for the patient to present recurrences [(3-5)]. In this respect it has been demonstrated a progressive reduction of chances of achieving gross total resection (GTR) with each subsequent resection [(6)]. More specifically, cranio cervical junction (CCJ) tumour localization has been associated with a poorer outcome and these patients should be managed in highly organized centers. As a matter of fact it is important to try to achieve complete resection in case of recurrences, but it must be stressed that surgical complications can severely reduce the patient's quality of life. So a maximal wise and safe resection has to be considered the best option in cases of recurrent chordoma, especially when dealing with patients in which realistic and curative expectations are lacking. In this paper we present our series of recurrent CCJ chordomas treated via a "tailored" approach, intending with this term, a combined multiportal, eventually multistaged, strategy. Personal considerations, technical aspects, outcomes and complications are extensively discussed.

Methods and Results

Case series

First case: recurrent CCJ chordoma with inferior extension to retropharyngeal/retrolaryngeal area and complete encasement of the left vertebral artery with subtotal erosion of the left-side of C1-C2 arch and dural sac compression. The patient underwent heavy ion radiotherapy. He was then treated surgically via a combined transoral-transcervical approach, after embolization of the left vertebral artery and craniocervical junction stabilization. Surgery was uneventful but post-operative the patient presented a major bleeding from tracheostomy that required accurate management in the OR. After that the patient presented a complete recovery with improvement of his symptoms in a few weeks. Adequate decompression of the spinal cord was obtained. Early post op MRI showed a very good decompression of the dural sac and an adequate resection of the tumour (Figure 1). Unfortunately the patient presented an aggressive re-recurrence after 4-5 months and a new surgical option was excluded. Currently, after 2 years, the patient is treated with palliation and chemotherapy.

Second case: recurrent CCJ chordoma with intradural involvement and extension to the right masticatory/parapharyngeal space. The patient had undergone previous radiotherapy. A two-day staged surgical procedure was planned and performed. Since the lack of viable local flaps due to the previous surgery, as a first step, a right-sided temporo-parietal fascial flap (TPFF) was raised for reconstruction. The right parapharyngeal compartment was then addressed via a transcervical submandibular endoscopic-assisted approach. The patient was fixed and finally, on day 2, the resection of the tumour was performed. A wide dural resection was created and reconstruction was done, with a multilayer technique, using fat, fascia and TPFF. Postoperatively a delayed cerebrosipinal fluid (CSF) leak occurred (after 3 weeks) with associated meningitis. The patient underwent revision surgery and was treated with antibiotics. After one year the patient is alive, with no actual evidence of recurrence and no swallowing problems. Post op MRI seems to show a gross total resection (GTR) (Figure 2).

Third case: recurrent CCJ chordoma with extensive involvement of the left parapharyngeal and extra-cranial jugular foramen areas after a transpetrosal and a transnasal approach. A one-day multiportal transnasal (left transpterygoid)-right transmaxillary endoscopic-assisted approach was performed. Extensive exposure of CCJ dura was obtained and the tumour seemed totally extradural. A right side nasoseptal flap was raised for recontruction. Surgery and post op were uneventful and patient has been subsequently treated with RT. The last MRI showed an adequate resection (Figure 3).

Fourth Case: recurrent CCJ chordoma after transnasal procedure. Tumour was located in both left and right parapharyngeal space, left petrous apex and middle cranial base. A staged surgery was planned. A combined transnasal transpterygoid and left transoral-transpharyngeal approach was performed to enable the management of left middle fossa, petrous apex and parapharyngeal compartments. A second stage was performed transorally to complete the resection of the parapharyngeal and upper spine lesions. The naso-septal flap was used to cover the left internal carotid artery (ICA) (paraclival and petrous portion). Surgery was uneventful. The patient has undergone postoperative radiotherapy. The last MRI showed a small stable remnant in the left petrous apex compartment (Figure 4). No major complaints are referred by the patient.

Discussion

When dealing with chordomas, two main issues should be taken into account: the need to obtain a GTR and the necessity to reduce at minimum post-operative morbidities and sequelae [7]. Recurrent chordomas represent in this scenario a unique challenge, especially when located in the cranio-cervical junction, given the extreme anatomical complexity of such region. Among recurrences, there is a big difference between cases, whenever recurrence occurs after exclusive surgery or eventually also after radiation therapy. Data from MD Anderson Cancer Center seem to demonstrate that in radiation-naïve patients repeated resections have a positive impact on local disease control. Furthermore, progression within 6 months from the last treatment is associated with a poorer outcome [1]. Technically speaking, in recurrent skull base chordomas, the limited ability to achieve a R0/R1 resection is related to the presence of scars that alter normal anatomical landmarks, hide embedded residual tumours and make surgical dissection more complex [2]. From a surgical point of view, the management of chordomas that extend laterally (also to the plane of the cranial nerves) and involve different anatomical compartments is challenging. In this respect, we absolutely agree with other authors [8] stating that these patients would benefit most from combined multiportal (one or multi-staged) approaches. Furthermore, in the setting of recurrences with an intradural extension, the topic of reconstruction can be really critical and challenging to manage due to previous surgeries and eventually RT. As a matter of fact, most of recurrent multi-compartmental lesions cannot be safely managed and adequately resected via a unique approach, and require a personalized strategy. This means using several ports (transnasal, transmaxillary, transoral, transpharyngeal, transcervical, transpetrous, etc), in combination or in staged procedures, and performing surgery both microscopically and/or endoscopically-assisted. There are several reports in literature of combined approaches to different areas of the skull base via multiple surgical stages [9]. Combination of transnasal and transoral approaches have been proposed to manage lesions with inferior extension [10]. In this respect endoscopic assistance seems to allow to overcome some limitations of traditional microscopic

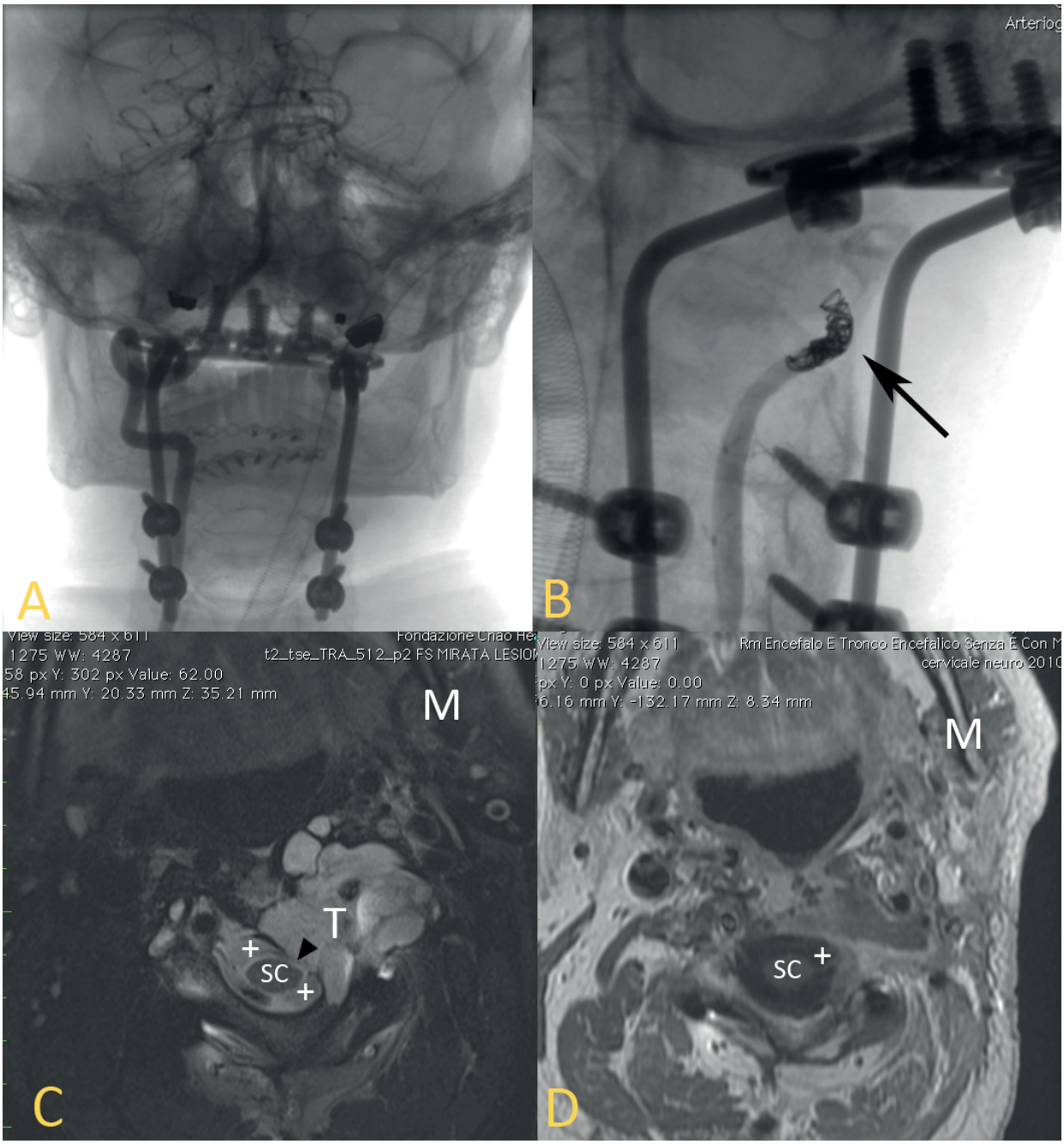

Figure 1. First case. A and B) preoperative angiography showing cervical junction stabilization and embolization of the left vertebral artery; embolization coil (black arrow). C) preoperative MRI, T2 weighted, axial plane, showing recurrent CCJ chordoma (T) with complete encasement of the left vertebral artery, subtotal erosion of the left-side of C1-C2 arch and dural sac (white crosses) compression (black arrow); spinal cord (SC), mandibula (M). D) early postoperative MRI, T1 weighted, showing decompression of the dural sac (white cross) and resection of the tumour; spinal cord (SC), mandibula (M).

transoral approaches. Another report describes the combination of a transnasal and far lateral, endoscopic-assisted, approach [11]. Our philosophy came from the universal lesson of Professor Draf "to not be dogmatic" and we have tried to apply this concept to every single case. It is well known that CCJ surgery can be associated with very severe complications [12] and it must be pointed out that the treatment itself can be potentially mutilating and disabling and have an unfavorable impact on patients' quality of life. In our series of recurrent CCJ chordomas in two patients we had a recurrence after exclusive surgery and in two after surgery and RT. The surgical strategy was planned and discussed exhaustively with the patients, as well as the expected results and outcomes that were explained in a clear and honest way to the patients. Surgical procedures, multistaged in two cases,

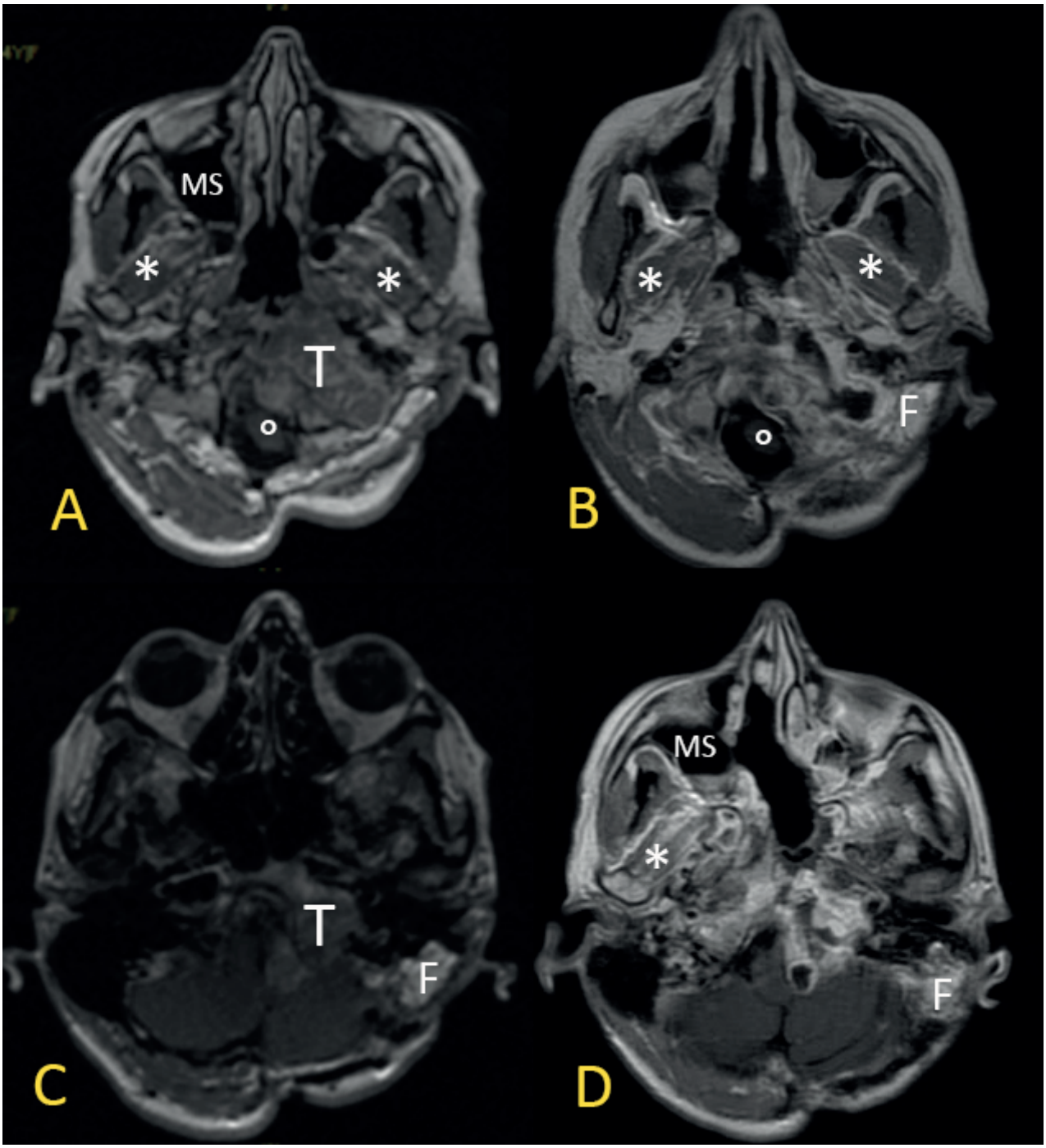

Figure 2. Second case. A and C) preoperative MRI, T1 weighted, showing recurrent CCJ chordoma (T) with intradural involvement and extension to the right masticatory/parapharyngeal space; maxillary sinus (MS), lateral pterygoid muscles (asterisks), spinal cord (white circle). B and D) postoperative MRI, T1 weighted, showing gross total resection of the tumour; autologous fat tissue from previous transpetrosal surgery (F).

always involved a multidisciplinary team (neurosurgeon-ENT). In two patients cranio-cervical stabilization was performed (in one case during the revision procedure planned as a 2-day surgery). A combined multiportal approach was performed, mixing different ports in all patients. As a matter of fact no patient was treated via the same procedure; for this reason we use the term of "treatment personalization". The two patients not previously irradiated were submitted to post op RT. There were no deaths related to the procedures in our series. At the time of writing, with a mean follow up of 28 months, all patients are alive, in 3 cases with persistent disease. In two out of three patients the residual disease seems to be stable while in one patient we are facing a devastating progression and chemotherapy and palliative support is currently being given. Complication rate was

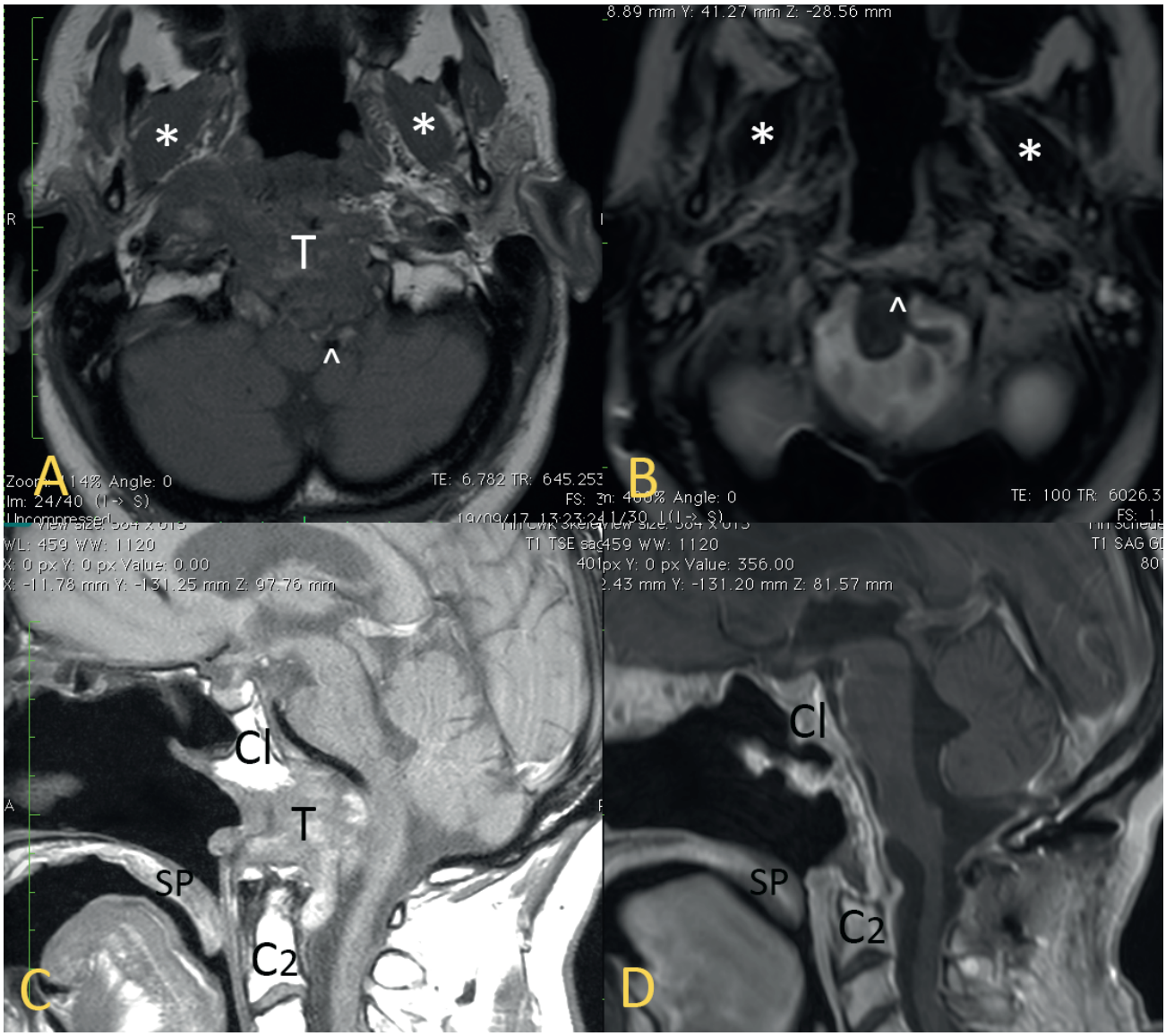

Figure 3. Third case. Preoperative MRI showing a recurrent CCJ chordoma (T) with extensive involvement of the left parapharyngeal space after a transpetrosal and a transnasal approach. A) T1 weighted, axial view; lateral pterygoid muscles (asterisks); basilar artery (white tip). C) T1 weighted, sagittal view; clivus (Cl), tumour (T), soft palate (SP), dens of axis (C2). Postoperative MRI showing adequate resection of the lesion. B) T2 weighted, axial view; left vertebral artery (white tip), lateral pterygoid muscles (asterisk). D) T1 weighted, sagittal view; clivus (Cl), soft palate (SP), dens of axis (C2).

acceptable. A post op cerebrospinal fluid leak and meningitis were observed in the patient with extensive intradural involvement. It should be noted though that this patient was previously irradiated and no local flap was available due to previous treatments. In one patient we had a post op bleeding from the tracheostomy, surgically treated in the OR. New neurological symptoms did not occur and no major swallowing problems were reported. As a matter of fact, the management of local failures can be extremely complex and it is clear that in some patients, due to recurrence extension, a pure midline approach, in a single procedure, may not be adequate, although endoscopic endonasal approaches have gained widespread acceptance and are nowadays considered the workhorse in most cases of clival and craniocervical junction chordomas [13]. In this respect, the modularity of a multiportal approach, with different angles of attack, allows the management of different compartments, not amenable to a single route. In other words trying to manage these multicompartimental lesions via an unique approach is associated with 2 main drawbacks: the first is related to the expected unsatisfactory resection of the lesion, and the second to the increased risks encountered when pushing into its own limits any given approach. Our experience of "personalized"

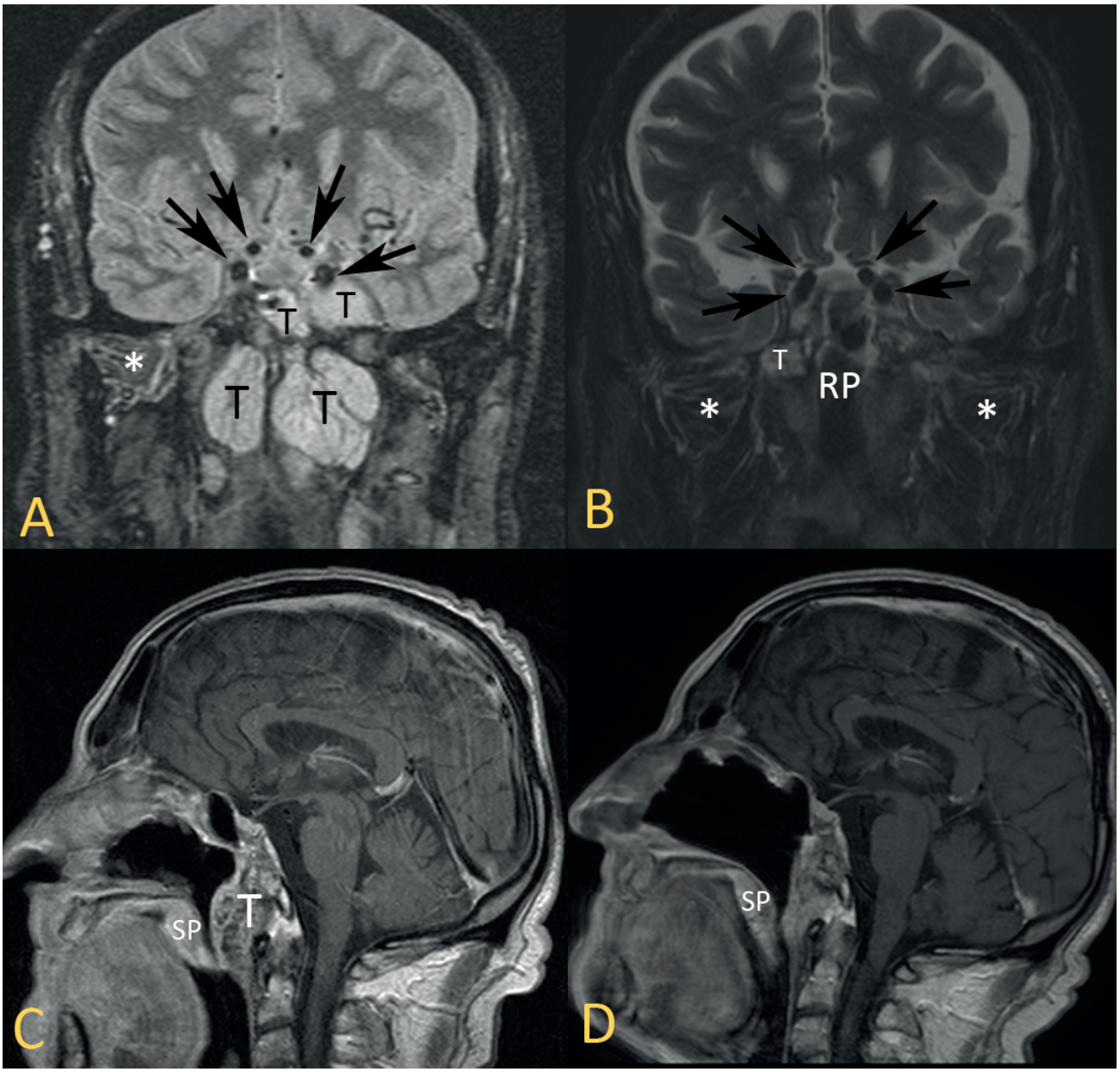

Figure 4. Fourth case. Preoperative MRI showing a recurrent CCJ chordoma after a previous transnasal procedure. A) T1 weighted, coronal plane, the tumour (T) is located on both sides in the parapharyngeal space, in the left petrous apex and at the middle cranial base; lateral pterygoid muscle in infratemporal fossa (asterisk), internal carotid artery (black arrows). C) sagittal view of the tumour (T); soft palate (SP). B) postoperative MRI, T2 weighted, coronal view, in which we can see a small stable remnant of the tumour (T) in the left petrous apex compartment; internal carotid artery (black arrows), rhinopharynx (RP), lateral pterygoid muscle (asterisk). D) sagittal view of postoperative MRI; soft palate (SP).

approaches could be considered a really interesting option for these complex cases. Obviously, anatomically speaking, different routes call for different landmarks. Summarizing the concept, when dealing with a transnasal approach to paramedian and lateral areas (petrous apex, jugular foramen) the key elements are represented by the vidian nerve, the eustachian tube and the foramen lacerum [14]. In this respect, the necessary removal of the pterygoid system opens the way to deeper areas. When coming through a transoral-transpharyngeal approach, whether or not endoscopically-assisted, the role of the superior constrictor muscle, styloglossus muscle and stylopharyngeus muscle is critical in guiding to the parapharyngeal portion of ICA [15]. Changing perspective, and coming through a lateral transcervical approach, the identification of the ICA can be done in the neck just below the sternocleidomastoid muscle and posterior belly of the digastric muscle. From there the artery can be followed to the upper parapharyngeal space, eventually by transecting the posterior belly of the digastric muscle and also the styloid ones

and the styloid process. It must be underlined that the ICA is always medial to styloid apparatus. When moving from a more lateral direction (tranpetrous approaches in general) the posterior vertical segment of the ICA can be found in close proximity to the cochlea and the bony part of the Eustachian tube (muscolotubal canal). The role of these structures cannot be underestimated. A different discussion must be done when dealing with the vertebral artery. This can be identified in a transcervical approach (antero-lateral approach) having in mind the anatomy of the suboccipital triangle and the role of the transvers process of C1. Once the control of the major vessels is achieved surgery can proceed more safely. We would like to underline the importance of a well-coordinated skull base team. With this concept we intend the presence also of dedicated anesthesiologists and care-givers. In other words, when dealing with these recurrences, one should keep in mind that an adequate and personalized surgical strategy is necessary but not enough and it is absolutely mandatory to work in a dedicated environment.

Our study has some obvious limitations. Among others, the number of cases, the retrospective nature and the short follow up period. On the other side, the rarity of the lesions and in particular of CCJ localizations makes, in our opinion, these data worthy to be known.

Conclusion

Although endoscopic endonasal approaches have gained widespread acceptance and are considered the workhorse in most cases of clival and craniocervical junction chordomas, in some cases of recurrence, or in presence of very lateralized lesions/ anatomical variations (i.e. aberrant course of parapharyngeal portion of ICA) midline approaches are either contraindicated or very difficult to perform. In all these cases it seems reasonable to consider a versatile strategy including different approaches, modulating the surgical needs with different answers and solutions offered by different routes. In other words to personalize as much as possible the approach, being creative and not dogmatic.

Authorship contribution

Each author has made substantial contributions to the work in terms of acquisition of data and all have approved the submitted version.

Conflict of interest

none declared.

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