Abstract: Objective: Presenting the first case of superior semi circular canal dehiscence syndrome in the Spanish literature and to establish, using embryological studies, the period in which superior semicircular canal dehiscence originates. Material and methods: 52 embryos and foetuses, from 6 mm to foetal maturity, were studied. The case of a patient suffering from superior semicircular canal dehiscence syndrome is presented. Results: The superior semicircular canal and the intracranial space are communicated through bony lacunae, in the period between the 24th and 28th week of foetal development, but this communication is discontinued in the 30th week. Permeability of these lacunae, later in life, could result in the development of superior semicircular canal dehiscence syndrome. The clinical results of the surgical repair of this abnormal communication, in this particular case, using a middle fossa approach and a transmastoid approach is presented. Conclusions: Superior Semicircular Canal Dehiscence Syndrome could be due to an abnormality in foetal development and its genesis, therefore, could be prenatal. Surgical repair via transmastoid approach is a reasonable alternative treatment to the middle fossa approach.

Key words: Dehiscence. Superior semicircular canal. Embryology Surgical approach.

INTRODUCTION

In 1998, Minor et al. reported 8 cases of patients suffering from vertigo and nystagmus in response to intense sounds or from pressure being exerted on the ear such as from Valsalva maneuvers or from swallowing; the majority of these patients also suffered from chronic imbalance. The clinical data together with the x-ray and surgical evidence from this study show that these 8 patients had ear dehiscence of the apical surface of the superior semicircular canal which explains the patients' symptoms. The dehiscent canal is in contact with the meningeal membrane that covers the middle fossa. This dehiscence is limited to the otic capsule and apparently does not affect the membranous labyrinth.

This, therefore, is a new syndrome that the authors of the study have identified. In regard to treatment for this new syndrome, Minor himself recommends surgery through a middle fossa approach to the lesion to achieve an improvement in and even full clinical recovery of the process.

This study presents a proven case of superior semicircular canal dehiscence syndrome (the first published in Spain) and an embryological study carried out on the superior semicircular canal in an attempt to try to identify the stage of the development involved in the genesis of the syndrome.

MATERIAL AND METHOD

In regard to histological material, we have studied 52 fetal embryos ranging from 6 mm to newborns belonging to the temporary collection of the Department of Human Anatomy and Histology of the Faculty of Medicine, Zaragoza, with the aim of studying the development of the superior semicircular canal and its relationship with the middle fossa. Following a careful dissection of the temporal bones, all the samples were preserved in 10% formol and decalcified with 1 to 3% nitric acid at a temperature of 25 degrees centigrade. After this process, the acid was eliminated by washing through the samples with running water. The temporal bones were dehydrated in alcohol at progressively increasing concentrations and were added to paraffin and cut into serial 7µm sections on a sagittal plane to the external auditory canal using a Leitz...
microtome. The preparations were stained using the Martins' trichromic technique.

Our clinical data is that of only one patient, but given the recent description of the syndrome (1998) and the lack of related publications in the Spanish language, we believe that is appropriate to include this case in our material. The patient was a 43 year-old male who had chronic imbalance as well as occasional vertigo on hearing certain intense sounds and during Valsalva maneuvers (such as blowing his nose and when defecating). He did not have either subjective hearing loss or tinnitus. The patient had been off work for more than a year. He had been diagnosed with anxiety and the origin of his vertigo labeled as psychogenic. The otoscopy was normal. The tone audiometry was normal, even though there was a 10dB gap in 250Hz (with bone conduction at -5dB). The patient suffered from the Tullio phenomenon when his right ear was stimulated with a 1500Hz tone at an intensity of 100dB; with Frenzel glasses a vertical-rotary nystagmus was detected, with an upwards slow phase rotating towards the left (clockwise). At the same time, the patient tended to displace his body towards the left. This same nystagmus and identical displacement could be detected when the patient received positive pressure on the external auditory canal using a Politzer bag, or when he performed a Valsalva maneuver. With suspected superior semicircular canal syndrome, a CT was done which confirmed the clinical diagnosis (Figure 5A).

The middle fossa was approached through a temporal craniectomy, lifting up the temporal lobe and exposing the arcuate eminence, in which a long, wide bony dehiscence could be seen, as well as a notable dehiscence of the tegmen in the tympani and antrum. We proceeded to close the dehiscence of the superior semicircular canal using a bone plate taken from the temporal craniectomy. The patient did not recover nor make any improvement following the intervention, continuing to have the same symptoms; a control CT revealed the displacement of the bone fragment that had been inserted and the persistence of the dehiscence. The patient agreed to undergo a second procedure which we carried out, but on this occasion through a transmastoid approach. A bone plate was taken from the compact bone of the mastoid surface and a wide mastoidectomy was performed along with skeletonization of the posterior and superior semicircular canals as far as the area of dehiscence. The abundance of pneumatized transalarirthine cells that passed through the superior semicircular canal was surprising. The dehiscence of the tegmen of the tympani and the antrum was also evident. The plane that existed between the hard and arcuate eminence was lifted and the bony plate of cortical mastoid was inserted between the two, using this same plate, duly manipulated, to close a good part of the tegmen dehiscence that had been widened by us in order to access the medial fossa. The mastoid was filled with abdominal fat.

RESULTS

Embryological study

A close relationship between the superior semicircular canal and the middle fossa through the meningeal structures exists during the embryological development of the superior semicircular canal; as can be observed in the 24 mm embryo (Figure 1). Initially this relationship is through the cartilaginous otic capsule, its thickness varying depending on the level of the cut studied. This fact can be appreciated in the embryos and fetuses, of less than 18 weeks of development, in which the superior semicircular canal is found in the interior of the cartilaginous otic capsule which is surrounded by the meningeal space.

Figure 1. 24 mm human embryo. Note the inside of the otic capsule from the cartilaginous structure to the superior semicircular canal (SSC) and its relation to the middle fossa (MF). Martins trichrome x 25.
The superior semicircular canal begins its ossification from two canalicular centers in week 19; the most apically centered one being the first to appear.

In the 24-week-old foetus, a continuity relationship can be seen between the meningeal surface and the perilymphatic space by means of the bony lacunae that surrounds the semicircular canal, (Figure 2).

In the 28-week-old foetus, the bony superior semicircular canal is already wrapped in compact trabeculae, even though bony lacunae that are still in contact with the meningeal space can still be seen (Figure 3).

From week 30, our observations indicated that these connections had disappeared and that the superior semicircular canal (SSC) is surrounded by bone. This bone suffers from remodeling, leaving - depending on the cut being studied - areas of greater or lesser thickness (Figure 4).

Results of the clinical case presented

The patient felt much better two months after the second intervention had been carried out; the Tullio phenomenon and the chronic imbalance having disappeared and the patient having re-incorporated himself back into normal working life.

In the control CT, (Figure 5B), the closure of the dehiscence of the canal and of part of the tegmen dehiscence can be seen.

DISCUSSION

More than 75 years ago, Tullio proved that the fenestration of a semicircular canal provokes nystagmus on the same plane as the open canal in response to intense sounds, because of which, this manifestation is today known as the Tullio phenomenon. The Tullio phenomenon-
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Figure 4. 32-week-old human fetus. We can see how the internal layer of the superior semicircular canal (SSC) is becoming more and more compact, the apical bony lacunae is thinner and narrower and the external layer which connects to the meningeal space (MS) is not fully formed. Martius trichrome x 25.

Figure 5A. Basal coronal CT of the right ear. Note how the superior semicircular canal is dehiscent in its apical region, leaving an abnormal connection in the arcuate eminence between the perilymphatic space and the intracranial space. The adjacent tegmen tympani is also dehiscent.

5B. Postoperative coronal CT of the right ear (transmatoid approach). A bony plate can be observed (fine arrows), taken from the cortical mastoid that extends over the arcuate eminence, obliterating the dehiscent canal and at the same time closing the dehiscent part of the tegmen. The surgical opening of the mastoid surface (wide arrow) can be seen in the form of a lack of bone. The cavity of the mastoidectomy (M) is filled in (abdominal fat is placed in the cavity to maintain the bony plate inserted between the meninges and the ear in place).

non was first of all associated with the erosion of the otic capsule caused by congenital syphilis3, later with adherence between the stapedial footplate and the saccule, then with a perilymphatic fistula and later with a diseased utricle in men4-7.

It has been calculated from post-mortem examinations that the percentage of cases with superior semicircular canal dehiscence in the superior part of the canal is approximately 0.5%,8,9, but it has not been proven that all the dehiscence in this area of the labyrinth has a clinical translation.

It has been suggested that the cause of this dehiscence is a postnatal change of the middle and external layers of the three layers that make up the canal roof9. Our results point in another direction given that we have found the alteration to be prenatal and not postnatal; we have obser-

ved a critical period in the development of the SSC, from 24 to 28 weeks in the embryological development, during which time a connection exists between the perilymph and meningeal spaces of the middle fossa. From the 28th week onwards, both spaces remain definitively separated by the formation of a fine bony layer that will belong to the internal peristeum9. In the 28-week-old fetus, we have not been able to observe the most external
The periosteum layer completely developed which in some areas is open to the meningeal space, nor of the middle layer that continues with big bony lacunae. At 32 weeks of development, our observations show how the intermediate area is disappearing, with the smaller surface of the bony lacunae, even though the external layer is still in contact in some areas with the meningeal spaces. This opening disappears at the end of the development, as we have been able to see in the 38-week-old fetuses. We believe that the ossification process of the superior semicircular canal is completed at the end of the development, a fact that differs from Carey’s observations, although we agree with this author that the ossification process of the temporal bone does not end until postnatal life.

If this connection were to persist, due to a failure in the subsequent bone reorganization of the otic capsule, it could take us to the origin of the superior semicircular canal dehiscence.

In the area of the disease and from an audiological point of view, the audiograms of the patients affected by the syndrome could be compatible with a mild conductive hearing loss given that a small gap between the air and bone conduction sometimes exists, especially at low frequencies. However, this “conductive hearing loss” is due more to a gain in conduction than to a real loss. This “increase in conduction” is not always evident and its presence is due to the fact that the existence of this third labyrinthine window creates some singular mechanical conditions in the labyrinth. In any case, the Weber test lateralizes to the dehiscent ear, but there is a normal stapedial reflex which enables a differential diagnosis for otosclerosis.

The following findings are from the vestibular tests of the patients affected by superior semicircular canal dehiscence syndrome, which should be evaluated using Frenzel glasses if dealing with nystagmic responses:

1. Vestibular nystagmus and vertigo induced by sounds. This happens in 88% of cases. The most evident responses are obtained sending the patient, via air conduction, sounds between 90 and 110dB between frequencies of 250 and 2000Hz.

2. Vestibular nystagmus and occasional imbalance of identical characteristics to that described in the previous section, in response to negative or positive pressure changes in the ear.

3. Other vestibular examinations that are more innovative, but that require specific stimulus and response recording equipment for them to be evaluated, are: the Vestibulo-Ocular Reflex (VOR) and the Vestibular Evoked Myogenic Potential (VEMP). Both examinations are based on the fact that the utricle and the saccule are located very near to the stapedial footplate which means that audible stimuli, such as clicks, can stimulate the sensorial endings of the corresponding macules, causing muscular (VEMP) or ocular reflex responses (ORR). These responses are made easier if a third labyrinthine window exists, as happens with dehiscence of the superior semicircular canal, given that the pressure exerted by the stapedial footplate is “diverted” in the direction of the posterior labyrinth in general and the utricle and the saccule in particular.

- Vestibulo-Ocular Reflex (VOR) evoked by high amplitude clicking sounds that appear below the normal threshold.
- Vestibular Evoked Myogenic Potential (VEMP) in response to clicking sounds that appears below normal thresholds and at higher than normal amplitudes.

From the diagnostic point of view, superior semicircular canal dehiscence syndrome is suspected when a patient has vertigo in reaction to intense sounds or to pressure changes in the ear frequently associated with chronic imbalance and confirmed with Computed Tomography. Other studies like the VOR and the VEMP are useful, but not definitive.

From the surgical point of view, the dehiscence is dealt with by closing it. In order to do this, the aim is to perform an intraluminal obliteration of the dehiscent part of the canal (plugging), then to cover the dehiscence with a bony plate. Another technique - resurfacing - consists of performing a simple closure of the dehiscence, repairing the surface with a bony plate. There are two surgical approaches to carry out these repairs: the middle fossa approach and the transmastoid approach.

In the clinical case presented, both approaches were carried out. First of all the middle fossa approach was used, proceeding to repair the dehiscent surface, plugging the dehiscence with a bony plate taken from a flake of temporal bone. The dehiscence did not close as we had intended it to due to the displacement of the bone graft during the repositioning of the retracted temporal bone and the more reasonable explanation for the failure is that the graft proved unstable. It is probably difficult to support a rigid bony plate on a curved surface such as the arcuate eminence, added to the restitution of the middle lobe to its anatomical position which is an additional destabilizing factor.

The transmastoid approach was appropriate for the repair carried out on the same patient six months after the previous surgical failure. It is necessary to know the surgical approach well in order to easily reach the area to be plugged, skeletonizing the superior semicircular canal while nearing the dehiscent zone directly and confidently at the same time that the tegmen is opened enough to guarantee direct middle fossa access from above and below. The placement of the bony plate that repairs the dehiscence is firm and stable, given that, on entry, the inserted cortical mastoid is left trapped between two surfaces that compress it and keep it in position.
In comparison, patients with superior semicircular canal dehiscence syndrome may have a dehiscence associated with the tegmen tympani and the antrum tegmen\textsuperscript{19}, which, to some degree, facilitates access to the apical region of the superior semicircular canal which, in our case study it was only necessary to widen a little. Because of this there have been reports that mastoid encephaloceles are associated with the dehiscence of the superior semicircular canal\textsuperscript{20}, due to an associated dehiscence of the tegmen. In these cases, the closure of the superior semicircular canal should be combined with repairing the tegmen so as to avoid cerebral hernias.

In conclusion, the genesis of dehiscence could take place in the prenatal period, agreeing with our preliminary results. In regard to which approach to use for the surgical repair, we found the transmastoid approach to be perfectly suitable to repair both the dehiscence of the canal as well as that of the tegmen, given that it allows for more stable repairs and has lower morbidity than the middle fossa approach.

References