EDUARD TOLOSA I COLOMER
TOLOSA-HUNT SYNDROME

The eponym

Tolosa-Hunt syndrome. A syndrome due to combined involvement of the common oculomotor, trochlear, trigeminal (first branch), and abducens nerves due to an aneurysm or thrombosis of the cavernous sinus and of the sella turcica that invades the external wall of the cavernous sinus[1].

Eduard Tolosa i Colomer
(1900-1981)

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Eduard Tolosa i Colomer was born in Barcelona on 24 October 1900. Graduating in medicine in 1921, he earned his PhD eight years later with the thesis *Algunas consideraciones sobre el diagnóstico diferencial de la enfermedad de Raynaud* (Some considerations on the differential diagnosis of Raynaud disease)[2]. Interested in internal medicine, he spent some time as an assistant at Ferrer i Solervicens' Medical Clinic, but soon became interested in studying neurological diseases. To that end, he worked for two years as an assistant in Barraquer i Roviralta neurology dispensary at the old *Hospital de la Santa Creu* and later with André Thomas at the *Hôpital Saint Joseph* in Paris. In France, he attended several of Guillain and Alajouanine's courses and also visited the departments headed by Foix, Lhermitte, and Barré. He completed his training with an internship in Madrid, where he worked in José Sanchís Banús's outpatient clinic. He also undertook an experimental study under the direction of Fernando de Castro in Gonzalo Rodríguez Lafora's department at the *Instituto Cajal*. Shortly afterward, he became interested in neurosurgery; in Tolosa's own words[2], due to "The wish to cure a significant group of neurological cases concerning which neurologists remained thoroughly impotent. Moreover, the direct experience of cerebral physiology and anatomy that neurosurgery provides led me irresistibly in that direction."

His first surgical interventions were conducted with Joan Puig-Sureda and in 1930, he began his neurosurgical studies at the *Hôpital de la Pitié* in Paris with Vincent and David. He would later visit many centres, among which Tolosa highlighted *Wenzel-Hanke Krankenhaus* in Breslau (nowadays Wroclaw), with Foerster, and the *Serafimerlasarettet* in Stockholm, with Olivecrona.

He was appointed associate professor of neurology at the *Universitat Autònoma de Barcelona* (1934), worked with Manuel Corachán i Llort

Eduard Tolosa i Colomer, 1960s

(perhaps the first Catalan neurosurgeon), was assistant neurosurgeon at Joan Puig-Sureda surgical clinic at *Hospital Clínic de Barcelona* (1935), and head of the Department of Neurology and Neurosurgery at Barcelona's *Institut Políclinic - Clínica Plató* (1934). But his major scientific and consulting activity began in 1940 when he was appointed director of the Department of Neurosurgery at the *Institut Neurològic Municipal de Barcelona*, founded four years earlier by Belarmino Rodríguez Arias[3], a responsibility he held until his retirement in 1970. He also worked as a neurosurgeon at the *Hospital de la Santa Creu i Sant Pau*, affiliated with Barraquer i Ferrer's dispensary (1944-1954), and at *Hospital del Sagrat Cor* and *Hospital de Nens Pobres* (1955). When in 1956 two departments of neurosurgery were created at what is today *Hospital Universitari Vall d'Hebron*, Tolosa was appointed to head one (Adolf Ley was appointed to head the other). The new department was organised with the help of Fuenmayor and Figuerola. In 1970, Tolosa retired and resigned from all his positions, although he would be appointed honorary professor at the Professional Neurosurgery School at the *Universitat Autònoma de Barcelona* in 1975. Eduard Tolosa died on 4 December 1981 after 17

years of fighting a disease he had treated many times: Parkinson disease[4].

Eduard Tolosa's intense scientific and consulting activity is reflected in nearly 200 publications. He is also remembered as a great clinician, and his contributions to both semiological and surgical aspects in the field of cancer and in the treatment of subdural and epidural hematomas merit special mention. He was a pioneer in stereotactic surgery in the 1960s, using it to treat Parkinson disease and to conduct cerebral biopsies[4]. Especially noteworthy is *Angiografía cerebral* (Cerebral angiography, 1953), one of the first publications in Spanish on this subject. He also penned *Síndromes extrapiramidals* (Extra-pyramidal syndromes) in *Monografies Mèdiques* (1931) and *Cirugía del dolor* (Surgery of pain, 1941). Moreover, he played an important role in scientific societies. He was a founding member of the *Sociedad Española de Neurología*, which he also served as treasurer and vice-president, and of the World Federation of Neurosurgical Societies. He was president of the *Sociedad Luso-Española de Neurocirugía* and of the *Associació de Neurologia i Psiquiatria de l'Acadèmia de Ciències Mèdiques de Catalunya i de Balears*[2-4].

To many, Tolosa, along with Adolf Ley, was the great initiator of Catalan neurosurgery and one of its earliest specialists in Spain. Eduard Tolosa and Sixto Obrador ("Dott-Obrador syndrome") are two Spanish neurosurgeons to have generated medical eponyms used worldwide.

Tolosa-Hunt syndrome

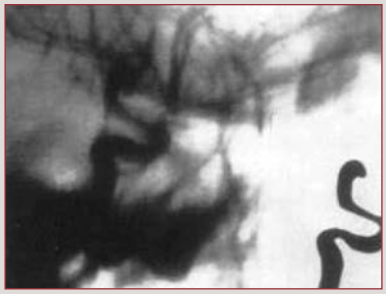
It is defined as unilateral ophthalmoplegia associated with retro-orbital or periorbital pain in the area innervated by the first branch of the trigeminal nerve. Attacks may alternate from one side to the other and may be accompanied or followed by paresis of isolated oculomotor nerves; alternatively, the full superior orbital fissure syndrome may appear.

Exceptionally, it affects the eye's sympathetic pupillary innervation. It is believed to be due to a non-specific inflammatory process and to granulomatous tissue in the superior orbital fissure or in the cavernous sinus[5]. It affects both sexes equally, and its onset is most common in the sixth decade of life. It is unilaterally localised, causing progressive retro-orbital pain and may manifest as a scintillating scotoma, sometimes recurring after remissions of months or years. Sometimes it progresses from blurry vision to total blindness. It may be due to diverse inflammatory processes of the cavernous sinus, and normally histopathologic study finds that the cavernous sinus is the only structure involved. Tolosa-Hunt syndrome is treated by treating its cause, although patients with granulomatous lesions improve with the administration of corticosteroids. The syndrome may last days or weeks; it may disappear spontaneously or after treatment, although it sometimes recurs[6].

This syndrome was first described in 1954 when Eduard Tolosa published a case report in *Journal of Neurology, Neurosurgery and Psychiatry*[7] that would immortalise him. Years later, he would describe it thus[2]:

> "In this study we reported the case of a 47-year-old man with a one-month history of pain in the left orbital region associated with left ophthalmoplegia that became complete within a week. Three years before, he had had a similar episode of some days' duration. Angiography of the left carotid ruled out an aneurysm of the infraclinoid carotid artery. Surgical exploration of the optochiasmatic region found no abnormalities. The patient died in the ward and the autopsy showed that the intracavernous portion of the left carotid siphon was wrapped in a sleeve of a cushiony tissue that encompassed the oculomotor nerves. Histological examination showed that this was nonspecific granulation tissue."

In 1961, Hunt and his collaborators[8] described six additional cases, employing their personal observations as well as clinical cases already



The image in the top shows the uniform narrowing of carotid siphon, whereas the image at the bottom is the granuloma wrapped around the carotid artery[7]

J. Neurol. Neurosurg. Psychiat., 1954, **17**, 300.

PERIARTERITIC LESIONS OF THE CAROTID SIPHON WITH THE CLINICAL FEATURES OF A CAROTID INFRACLINOIDAL ANEURYSM

BY

EDUARDO TOLOSA

From the Neurosurgical Service of the Neurological Institute, Barcelona, Spain

The clinical diagnosis of subclinoid carotid aneurysms is generally based on the characteristic association of two features : (*a*) pain in the first division of the trigeminal nerve ; and (*b*) progressive paralysis, partial or total, of the oculomotor nerve and occasionally of the fourth, sixth, and the fifth cranial nerves. Although the symptomatic association just described is highly characteristic, it is only through the use of cerebral angiography that we are able to establish the diagnosis with certainty.

In some cases of the syndrome, arteriography may indicate the presence of a tumour in the base of the skull (the carotid siphon running a zigzag course or showing an abnormal opening of its second curve, etc.), but in others there may be a non-aneurysmal vascular process of the intracranial carotid. In this paper I wish to deal with the latter possibility, and to report a case in which the evolution and clinical condition of the patient seemed to point to the diagnosis of intracavernous carotid aneurysm, while the arteriographic examination suggested the existence of disease of the wall of the carotid siphon, and this was subsequently confirmed at necropsy.

Case Report

S. M. M., a man aged 47, was admitted to the Neurological Service of the Neurological Institute in September, 1949. His present illness began three years before, the initial symptom being pain in the left orbital region. The ache was slight but continuous for a few days. A month before the patient's admission pain was again experienced in the territory of the first branch of the left trigeminus : it became violent and continuous, and was accompanied by vomiting. During the past eight days the patient had noticed ptosis of the upper left eyelid which increased rapidly into a total left ophthalmoplegia.

Funduscopic examination revealed no important pathological changes. The visual fields were full. Visual acuity : right eye, 1 ; left eye, 0·5.

The third, fourth, and sixth cranial nerves on the left side were completely paralysed. The left corneal reflex was weak. Objective sensibility was normal in the region of the left fifth nerve. No disturbances of the trigeminal motor functions or defects in the remaining cranial nerves were found. The neurological examination of the trunk and limbs was also negative. Blood pressure was 140/70 mm. Hg.

Wassermann's test in the blood was negative.

The radiological examination of the skull showed only an increase in density of the small sphenoidal wing on the left side, the upper limit of which appeared ill-defined.

In the angiographic pictures, the anterior and the middle cerebral arteries appeared in their normal positions but the carotid siphon showed in the lateral view (Fig. 1) a segmentary narrowing at the level of portion C_2.

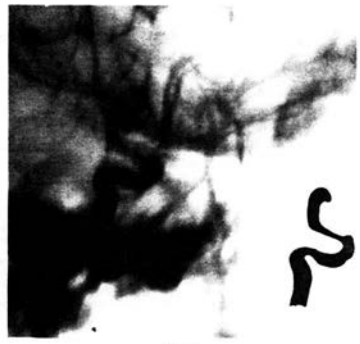


FIG. 1

This examination apparently excluded the diagnosis of saccular aneurysm or of meningioma. The existence of a malignant tumour in the base of the skull seemed

Reproduction of the first page of Eduard Tolosa's article in *Journal of Neurology, Neurosurgery and Psychiatry*[7]

published under the general denomination of painful ophtalmoplegia. They attempted to describe a new syndrome basically following the criteria of periorbital pain, ophthalmoplegia, trigeminal nerve dysfunction, frequent spontaneous remission and recurrence, and lack of involvement of other structures. Hunt asked Tolosa for his histological preparations[2], which enabled him to prove the existence of nonspecific granulation tissue within the cavernous sinus, with no primary arteritis. Tolosa published another case with spontaneous remission in 1961[9]. Then, five years later, Smith and Taxdall[10] published four new cases, designating the new anatomo-clinical entity Tolosa-Hunt syndrome, adding the response to corticosteroids as an additional diagnostic criterion.

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