Medicina Historica 2020; Vol. 4, N. 1: 35-38

A case of juvenile nasopharyngeal angiofibroma belonging to the Pathology Museum of Turin

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Abstract. The Pathology Museum of the University of Turin houses historical dry and wet specimens dating back to the end of XIX and begin of XX century. Among these specimens, a case of juvenile nasopharyngeal angiofibroma was discovered during the study of the diagnostic re-evaluation of the Collection. Juvenile nasopharyngeal angiofibroma is a rare non-capsulated vascular neoplasm predominantly arising in adolescent males. It suggests that this neoplasia could be androgen dependent, but this hypothesis remains controversial and its origin is still not clear. It is considered a benign neoplasm but locally aggressive and it could spread to the base of the skull and into the cranium. The museum wet-specimen dates back to March 31th 1911, as shown on the autopsy report which number is reported on the original label on the jar. The specimen was collected performing the autopsy on a 14years old boy who died of unknown reasons, most likely for epistaxis or suffocation. The only pathological finding is the nasopharyngeal lesion. Macroscopic examination of the specimen shows that on the base of the skull there is a huge polypoid lesion. Histopathology shows an edematous fibrous stroma and staghorn blood vessels irregular in size and shape. The tissue appears to be well preserved despite the long extended period in fluid. The original diagnosis was "Nasopharyngeal polyp" and it was actually the name of juvenile nasopharyngeal angiofibroma before the first official report of this lesion in the year 1948.

Key words: pathology museum, nasopharyngeal angiofibroma

Introduction

The Pathology Museum of Turin has an ancient history since their foundation dates back to 1818. Doctor Giovanni Pietro Gallo started to collect the most particular pathological specimens founded performing autopsies. In following years, the collection grew and in 1872 there were more than 1000 specimens (1). When Professor Pio Foà became Director of the Institute of Pathology, a new kind of collection started: not the strangest specimens like a Wunderkammer, but specimens with a scientific interest, although not so spectacular. The director especially collected neoplasms and infective diseases. His follower Professor Ferruccio Vanzetti was instead interested in cardiovascular pathology (2) and collected many specimens of tertiary syphilis. Nowadays, the Pathology Museum houses 304 wet specimens representative of many neoplastic and infective diseases. In recent years, a project of diagnostic re-evaluation of the specimens with modern techniques of pathology has started. This project includes the sampling of the specimens with a conservative approach respecting the macroscopic integrity of the specimens (3). Core biopsy is the technique of choice in the huge masses without sections already present, whereas the old sections, can be useful for new sampling without damaging the specimens. Then the study of the sampled tissue is carried out using histochemistry, immunohistochemistry and if possible molecular biology as well. This study allows the diagnostic re-evaluation of the specimens starting from the original diagnosis on the label. The

old diagnosis are not often so easy to understand for a modern pathologist. Therefore old books of Pathology are used for a better comprehension. The most useful book is the text of Pio Foà. Professor Foà wanted Italy to have a Pathology book in its own language, until then there were no any books from Italian authors. The book "Trattato di Anatomia Patologica" dates back to 1921 (4). Pio Foà personally wrote the chapters of haematology and respiratory system. This book helps to understand the old diagnosis especially when they use terms that now are not so clear according to the modern classification of pathology. The integration between autopsy report, histology of the specimen and bibliography allows a correct diagnostic re-evaluation of these old wet specimens.

Material and methods

Among the specimens of the collection of the Pathology Museum of Turin a case of "nasopharyngeal polyp" has been re-evaluated. This diagnosis is reported on the original label (Fig. 1) together with the number of autopsy. The autopsy was performed on March 1911. Its number is 12238. It was the case of a 14years old boy. There is not a certain cause for the death, but there is the description of a huge mass "as big as an egg" on the base of the skull. No other pathological findings are reported. The jar appears in poor condition: the fluid is reduced and the specimens is not fully covered. The fluid shows a dark brown colour and it is impossibile to see the specimen inside the jar (Fig. 2). Surely, this specimen needs to be restored in a future. Despite these bad conditions, the wet specimen shows an excellent preservation, probably due to the first fixation. Inside the jar there are two halves of a lower skull. The left half exhibits a lesion on the base of the skull which occupies the nasal cavities, therefore a possible cause of death of this boy was suffocation (Fig. 3). The lesion is grey and soft and there is no evidence of a capsule, even if it is well demarcated. A small sample was taken from a hidden surface of the lesion without damaging the specimen. Haematoxylin-Eosin stain was performed for a first evaluation of the morphology.

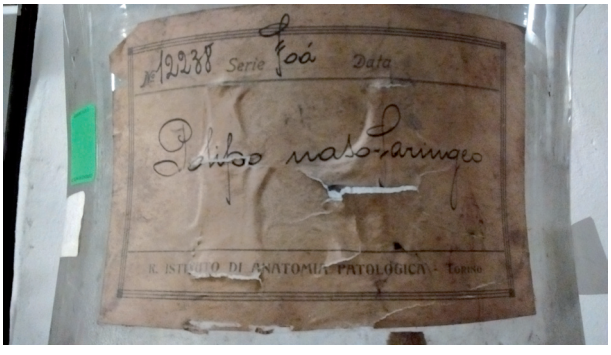

Figure 1. Original label on the jar

Results

Histopathology shows a loose edematous stroma with few cells interspersed with many staghorn

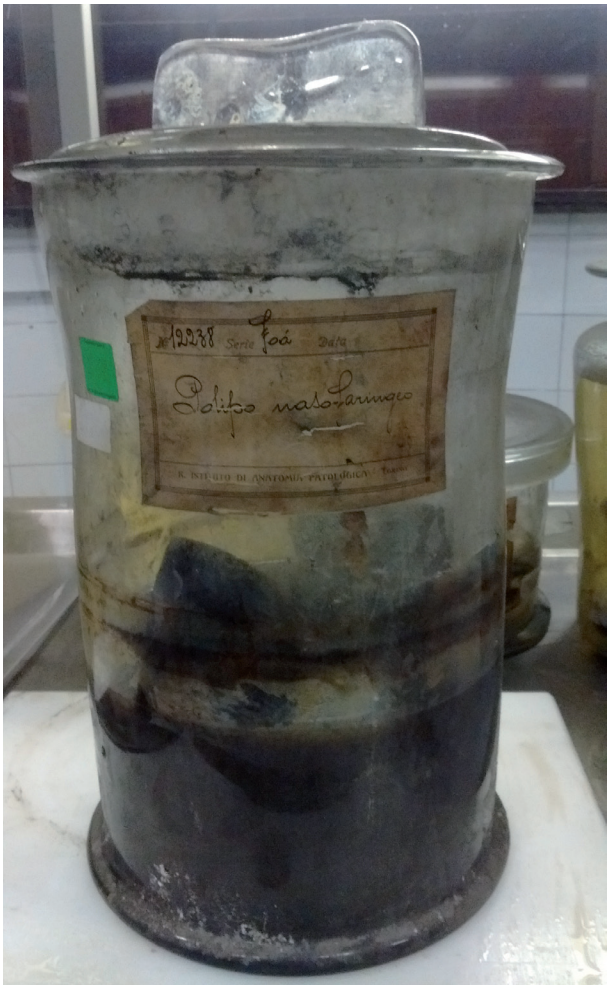

Figure 2. The jar in original conditions

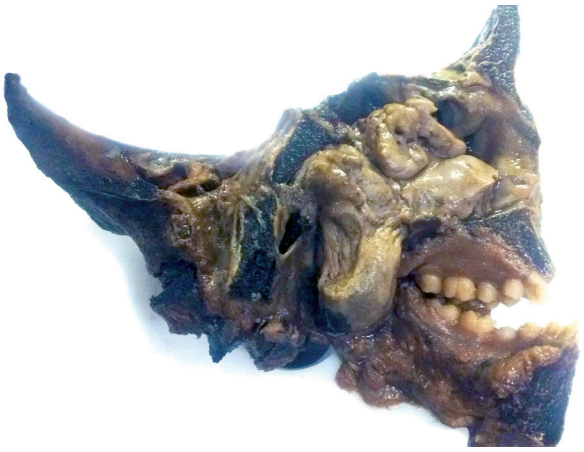

Figure 3. The wet specimen

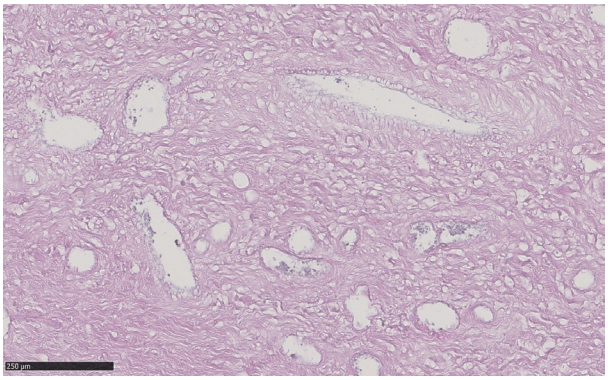

Figure 4. Pathological section H&E stain 10x magnification

blood vessels irregular in size and shape (Fig. 4). The morphological preservation is good despite the poor conditions of the jar. The morphology of the lesion is pathognomonic for nasopharyngeal angiofibroma. Therefore the diagnosis does not require further specific investigations such as histochemistry or immunohistochemistry.

Discussion

Juvenile nasopharyngeal angiofibroma is a very uncommon neoplasm, usually unilateral. It is benign but locally aggressive and it could spread to the base of the skull and into the cranium (5).Usually this lesion is located on the superior edge of the sphenopalatine foramen, but any other localisation in nasal cavity and nasopharynx is possible as well. The clinical presentation is very typical: a nasal obstruction with epistaxis, rhinorrhea and pain. Nowadays these lesions are diagnosed by MRI and angiography and early diagnosis is required for an early treatment. An early diagnosis allows a combination of preoperative embolization and surgical resection and the prognosis is good (6). The innocuous presentation of symptoms often delays the diagnosis and advanced lesions are more difficult to treat. This lesion affects typically male between 14 and 25 years of age, probably due to the high expression of androgen receptor of this neoplasm. Some authors suggest hamartoma and malformation theory, but the origin of juvenile nasopharyngeal angiofibroma is still not understood. Historically Greek, Roman and Arabian doctors used the term nasal polyps for all the masses within the nasal cavities or nasopharynx. In year 1847, Chelius wrote "fibrous nasal polyps commonly occur in person about the time of puberty" (7). Surgical texts of the early XIX century describe often how to remove these bulky lesions which could produce a typical face known as "frog face". Legouest in 1865 noted the predilection of these lesions for young male (8) and Gosselin in 1876 (9) noted the possibility of spontaneous regression after puberty. Chaveau in 1906 suggested the name of juvenile nasopharyngeal fibroma (10), Friedberg in 1940 first used the term angiofibroma (11). The first article in literature about juvenile nasopharyngeal angiofibroma dates back to 1948 (12). Therefore the diagnosis of "Nasopharyngeal polyp" on the label was correct according to the knowledge of the time. This is the second museum specimen showing this disease reported in medical literature. The other case was that of a specimen of a maxillectomy performed in 1841, which was diagnostically re-evaluated in 1987 (13).

Conclusion

This case shows a rare neoplasm that killed a young boy in absence of surgical treatment. This specimen shows the spread of this lesion demonstrating how it can evolve in its natural history and how this lesion can be lethal even if it is biologically benign. Probably the boy died for suffocation but an epistaxis is possible too. Unfortunately, the autopsy report can not help to understand the final cause of death. This specimen increases the importance of wet pathological specimens as historical heritage and biological archive as well.

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