

HISTORY OF ONCOLOGY

The distinguished Austrian ophthalmologist Ernst Fuchs (1851-1930) and the “sarcom des uvealtractus”

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Summary

Born in Vienna, in mid 19th century, Ernst Fuchs became an innovative ophthalmologist and is currently considered as the father of modern ophthalmology. Conditions like heterochromic uveitis and endothelial dystrophy were discovered by Fuchs and a series of ocular diseases are named after him. His work dealing with eye pathology, entitled: “Textbook of Ophthalmology”, became known as the masterpiece of the 19th century ophthalmology. In ocular oncology, Fuchs in-

roduced first the “sarcom des uvealtractus” (uveal sarcoma), known today as uveal melanoma. His treatment approach by enucleation was in vogue for more than 100 years, while his theory for hepatic metastasis of ocular cancer remains valid.

Key words: Ernst Fuchs, history of oncology, ocular oncology, uveal melanoma

Introduction

Cancer affected mankind since prehistoric times and it was recognized as a distinct entity since the Hippocratic era [1,2]. Ocular and orbital wall cancers were mentioned in the work of the ancient Greek medico-philosophers and were considered incurable, lethal, and non-operable malignant entities. Paul of Aegina (ca 625-690 AD) was the first to refer to this type of cancer and proposed non-invasive palliative measures [3]. Later on, in a series of ophthalmologic treatises of the medieval period, Arabo-Islamic physicians such as al-Mawsi-li (9th-10th century), al-Kahhal (ca. 940-1010), Haly

Abbas (10th century) and al-Sadili (14th century) provided several references concerning eye cancer focusing on eyelid tumours and cancerous ulcers of the cornea [4].

Moreover, various theories paved the way for the scientific research in oncology and ocular oncology too. Firstly it was the excess of humours (especially that of black bile) which was introduced by the ancient Greeks to interpret cancer [2]. Then, in 19th century, the germ theory of cancer, proposed that microorganisms were at the origin of the disease [5]. However, it was the invention of the



Figure 1. Portrait of Ernst Fuchs, 1910.

ophthalmoscope by the German physician and physicist Hermann Ludwig Ferdinand von Helmholtz (1821-1894) in Heidelberg during 1851, alongside with the concept of cellular pathology as developed by German physician and biologist Rudolph Virchow (1821-1902) in Berlin during 1858, which enabled the early understanding of intraocular tumours. At the same period, the development of general anaesthesia allowed the widespread use of various surgical treatments, including enucleation [6,7].

Among the pioneers in the field of ocular oncology stands Ernst Fuchs, whose expertise came from his ability to comprehend critical and pathologic features and from his extremely busy clinical practice (Figure 1). Living in a period of medical revolution, Fuchs was able to correlate the ophthalmic appearance and histopathologic features of the enucleated eye, as he did in his description of “sarcom des uvealtractus” (uveal melanoma) [7].

Ernst Fuchs’ life and work

Ernst Fuchs was born in Vienna on November 14, 1851, the eldest of three children, originating from a family of humble origin. In 1860, Fuchs enrolled at Scott’s Gymnasium in Vienna and graduated eight years later, in 1868. Despite his father wish to become an engineer, Fuchs decided to follow medicine. During the period of his studies, he

had the chance to be taught by authorities in the medical field like the anatomist Joseph Hyrtl (1810-1894), the physiologist Ernst Wilhelm von Brucke (1819-1892), the pathologist Carl von Rokitsky (1804-1878), the dermatologist Joseph Skoda (1805-1881), the surgeon Christian Albert Theodor Billroth (1829-1894), and the ophthalmologist Carl Ferdinand Ritter von Arlt (1812-1887) who became his mentors and worked together for several years [8,9].

In 1881, Fuchs became professor of ophthalmology at Liege, Belgium, the youngest Austrian-trained graduate to become a professor. In 1885, he returned to Vienna, succeeding Eduard Jaeger Ritter von Jaxthal (1818-1884) as clinical director at “Vienna eye hospital” which later became known as “Eye clinic of Fuchs” (Figure 2). He remained at this position until his retirement, in 1916. Under the administration of Fuchs, the clinic became a European centre of ophthalmology, where young physicians were trained and inspired by Fuch’s teaching [9-11].

Fuchs became a member of 39 European scientific societies and he was fluent in English, French and Italian and basic Latin and Greek [12]. Multifaceted personality, he loved literature, art, history, botany and geography. He also traveled a lot around the globe as a member of the Vienna geographical society [8]. It is said, that after his trip in Scandinavia, he brought with him “snow shoes”, introducing skiing in Vienna [13].

His masterpiece “Textbook of Ophthalmology”, was first published in 1889 and was regarded as the “bible of ophthalmology” for more than 50 years. It was originally written for his pupils in order not to be distracted by writing notes during his lectures. However, it became a classic book for ophthalmologists, translated into various languages, including Russian, English, Chinese and Japanese and its last edition in German appeared in 1945 [8,14].

Fuchs, was an excellent clinical observer who first described a series of eye conditions. Capable microscopist, he explained and categorized several pathological ocular lesions which bear his name, such as Fuchs’ endothelial dystrophy, Fuchs’ heterochromic cyclitis, Dalen-Fuchs’ nodules and Fuchs’ coloboma [8,10]. In 1906 Fuchs described a chronic, unilateral iridocyclitis that was characterized by iris heterochromia, naming it “heterochromic uveitis” or “uveitis syndrome” [15]. In 1910, he reported first a series 13 cases of central corneal clouding, loss of corneal sensation and formation of epithelial bullae; he named this condition “dystrophia epithelialis corneae” [16]. Fuchs never gave up research and even after his retirement he published 99 scientific manuscripts [8].



Figure 2. Ten Viennese doctors among which Ernst Fuchs, Process print, 1913.

Fuchs' description of uveal melanoma

Uveal melanoma is a rare malignancy, which arises from melanocytes within the uveal tract of the eye. It depicts the most common primary intraocular cancerous tumour in adults, representing



Figure 3. Uveal Melanoma, Laird Harrison, Medscape, 2017.

3-5% of all melanomas (Figure 3). Significant visual morbidity and metastatic progression is not uncommon, portending an extremely poor prognosis. Despite the excellent rates of local disease control with surgery or radiotherapy, around 50% of the patients will ultimately develop metastatic disease. The most common initial sites for metastases are the liver (60.5%), the lung (24.4%), the skin/soft tissue (10.9%), and finally the bones (8.4%). The overall rate of survival from initial diagnosis is between 51-69%, but in metastatic cases, this is lower than 8% [17,18].

In 1882, Fuchs described first the diffuse malignant melanoma of the uveal tract, introducing the most common primary intraocular neoplasm in adults, naming it “sarcom des uveal tractus” (uveal sarcoma), known today as “uveal melanoma”. According to his opinion, sarcoma of the uvea may proliferate directly from a highly developed type of uveal cell. He mentioned that patients with uveal sarcoma (melanoma) almost inevitably die and that enucleation (removal of the eye that leaves the eye muscles and remaining orbital contents intact) was the treatment of choice [19]. Using ophthalmoscope and microscope, he described 14 types of uveal sarcoma such as spindle and round celled sarcoma, sarcoma with areolar pigmentation, endothelial sarcoma, cavernous sarcoma, fibro-sarcoma, alveolar sarcoma, giant-celled sarcoma and

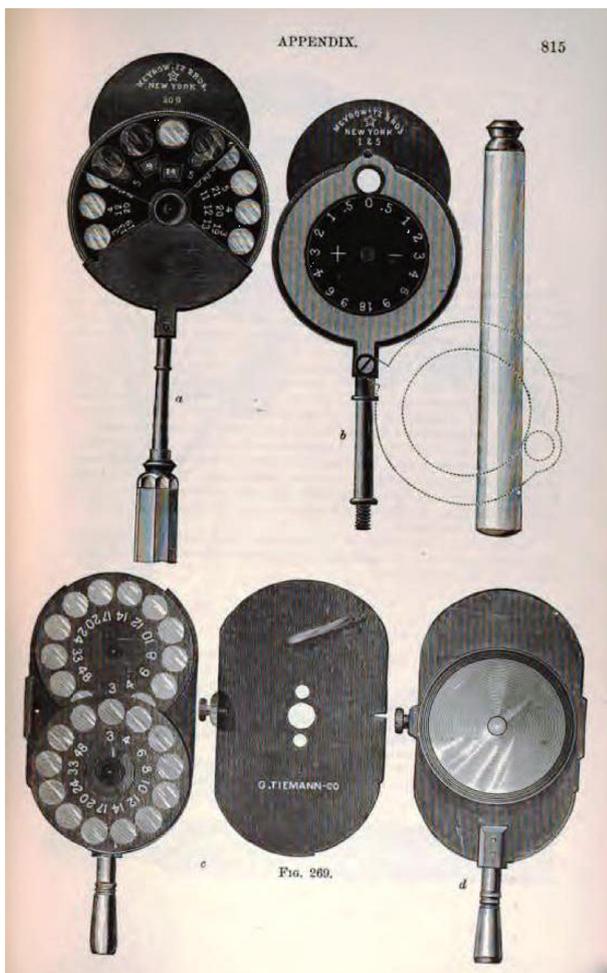


Figure 4. The ophthalmoscope that has been used by Fuchs, Textbook of Ophthalmology, 1882.

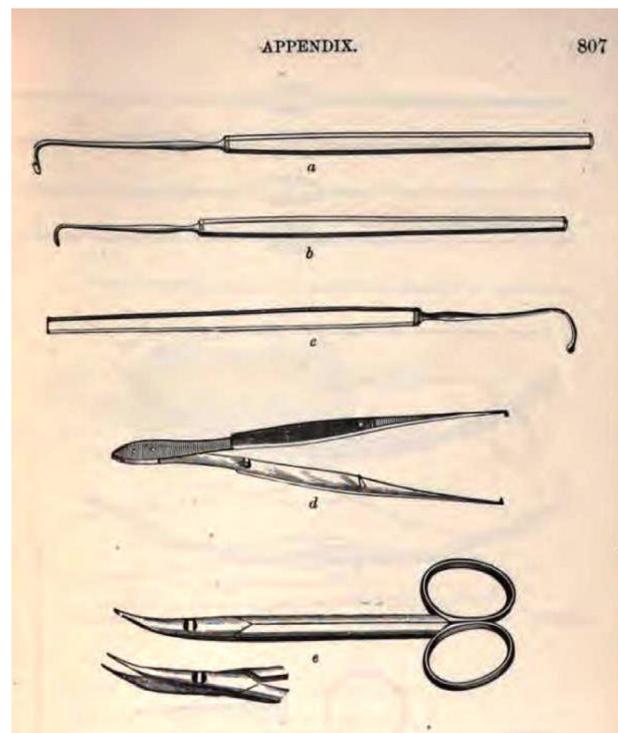


Figure 5. The surgical instruments that have been used by Fuchs for the operation of enucleation, Textbook of Ophthalmology, 1882.

myxosarcoma [20] (Figure 4). He also realised and described first the concept of metastatic tropism of the uveal tract sarcomas to the liver [21].

Discussion on Fuchs' "sarcom des uvealtractus"

Ernst Fuchs supported that ocular pathology belongs to ophthalmologists. Ocular oncology was for him a new challenge. Although he had understood that most sarcomas of the uveal tract contain more or less pigment, he avoided to use the term "melanoma". He stood by the old nomenclature, "sarcoma", as he did not believe that the normal chromatophores (melanophores) were derived from the neoplasm. Also, he observed that choroidal tumors usually began in the outer layers of the choroid [19]. Fuchs proposed that sarcomas should be treated as soon as possible by enucleation, a method which became the gold standard for a century [19] (Figure 5). In 1970s, a series of studies conducted by Zimmerman, McLean and Foster introduced a new therapeutic approach [22]. In three

manuscripts, the authors put forward a hypothesis which doubts enucleation as a treatment of the primary tumour, developing alternative therapeutic modalities such as radiotherapy and tumour resection [23-25].

Conclusion

Ernst Fuchs died in Vienna on November 21, 1930 after a brief illness of angina pectoris. After his death, he was described as "a man of great simplicity of character, of great industry and with a remarkable interest in all subjects; he not only made many valuable contributions to his specialty, but has helped and inspired pupils who are scattered all over this world" [10]. Fuchs' "Textbook of Ophthalmology" is considered to be the most influential 19th century monograph, while his paper about malignant sarcomas had a huge impact for more than 100 years. He is thought as the father of modern ophthalmology, the ophthalmologist who paved the way for future research on uveal melanoma.

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