B-ENT, 2014, **10**, 319-323

Historical Case: "Portrait of a One-Eyed Man" by Vincent Van Gogh

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Key-words. Neurofibromatosis type I; frontal sinus osteoma; frontal sinus mucocoele

Abstract. *Historical Case: "Portrait of a One-Eyed Man" by Vincent Van Gogh*. The post-impressionist Dutch painter Vincent Willem Van Gogh (1853-1890) painted the "Portrait of a One-Eyed Man" (1889) when he was admitted to the mental hospital of Saint Paul-de-Mausole. The portrait probably depicts one of Van Gogh's fellow patients who was suffering from a left-sided upper eyelid ptosis. Neurofibromatosis type I with orbitotemporal involvement has been suggested as the underlying disease process. However, from an otorhinolaryngological point of view, alternative diagnoses are possible. In this paper, the entities of giant frontal sinus osteoma and giant frontal sinus mucocoele are discussed, as well as the operative procedures available at the end of the nineteenth century to treat these lesions.

Introduction

The post-impressionist Dutch painter Vincent Willem Van Gogh (30 March 1853-29 July 1890) is well-known because of his unique artistic style and subsequent substantial and far-reaching influence on the development of art in the twentieth century. His personal artistic signature and the abundant availability of original information about his life and work (all the preserved letters to and from friends and family) made Van Gogh one of the most famous painters of the nineteenth century. However, during his career, Van Gogh's work was known to only a handful of people and was appreciated by fewer still. He suffered for years from mental illness, culminating in an incident in which he cut off part of his left ear on 23 December 1888 in Arles after a quarrel with his friend and fellow-painter Paul Gauguin. After this dramatic event, he committed himself on 8 May 1889 to the mental institution of Saint Paul-de-Mausole, a former monastery in Saint-Rémy, near Arles.[1] Since Van Gogh also wanted to paint portraits in Saint Paul-de-Mausole, he was dependent on the willingness of his fellow inhabitants to pose for portraits. During his stay in Saint Paul-de-Mausole, he painted a portrait of one of his fellow patients. He wrote about this painting in a letter to his mother in October 1889: "I am working on a portrait of one of the inmates here. It is strange that, when one has been here for a time and is used to it, one no longer thinks they are crazy".[2] It is quite likely that Van Gogh was referring to the portrait of a man with a deformed face which became known as "Portrait of a One-Eyed Man" (Figure 1). With his collar turned up, his hat tilted backwards and a cigarette in his mouth, the subject makes no obvious impression of insanity. On the other hand, Van Gogh has emphasised the ugliness of the man by accentuating the curvature of the injured eye with additional dark lines, rather than idealising him by focusing primarily on his "normal" side. The original colouring of the painting is difficult to guess at because the unpainted white areas of the unprimed canvas are now coloured brown because of the wax used when the work was relined.[3,4] At the moment, the painting is part of the collection of the Van Gogh Museum in Amsterdam (The Netherlands).

The Portrait of a One-Eyed Man: neurofibromatosis type I?

The painting shows the face and upper body of a man smoking a cigarette who is seen from his right. A left-sided upper eyelid ptosis is visible, and the left upper eyelid also has an oedematous appearance.

Disclosure: The authors have no financial interest in relation to the content of this article.

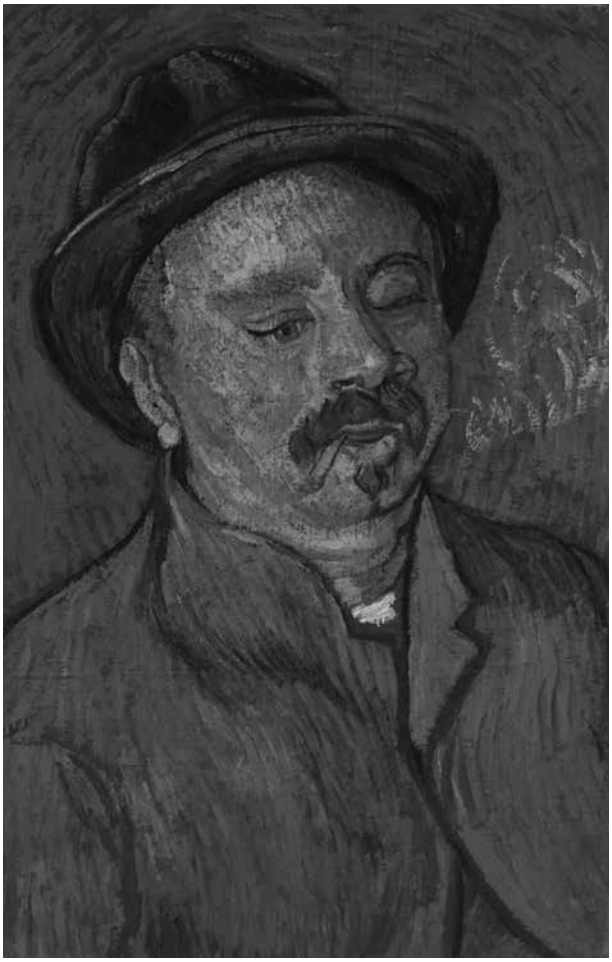

Figure 1
Portrait of a One-Eyed Man by V. Van Gogh (1889). *With courtesy of the Van Gogh Museum, Amsterdam, the Netherlands.*

The eyelid is painted with broad brushstrokes and so it is difficult to see whether the eyelid is oedematous or not. There is a suspicion of oedema given the reddish look of the left upper eyelid. However, the oedema would not seem to be sufficiently prominent to account for the complete closure of the eye, which also suggests upper eyelid ptosis as an important and independent factor. The left eyebrow also seems to be discretely pushed downwards (brow ptosis), suggesting a swollen left frontal region, with the mass effect deforming the eyebrow. The man is painted with Van Gogh's characteristic firm brushstrokes and so a frontal swelling cannot actually be observed. The lower eyelid seems not to be involved in the process. The left temporal region is not visible and the rest of the face seems to be normal.

In his book "The artist and the doctor", which discusses medical conditions and pathologies depicted in classical and modern paintings, Professor E.J. Dequeker suggested that Van Gogh's fellow patient had possibly been suffering from neurofibromatosis.[5] Neurofibromatosis type I (NF1) or Von Recklinghausen disease is an autosomal dominant disorder found in 1 in 3000 to 4000 live births, with an incidence of head and neck involvement ranging from 1% to 22%.[6] Affected patients may present with orbitotemporal deformities characterised by cutaneous plexiform neurofibromas of the eyelid and/or orbital involvement (possibly in combination with anomalous development of the sphenoid bone) in less than 1% of cases.[7] Plexiform neurofibroma of the eyelid predominantly involves the upper eyelid, manifesting as an S-shaped mechanical ptosis of the upper eyelid.[8] Ptosis of the brow region due to brow infiltration is uncommon, as suggested in the series of Lee *et al*.[6] However, looking closely at the depiction of Van Gogh's fellow patient, the S-shaped ptosis with preserved brow height which is typical for NF1 cannot be seen. Since eyelid ptosis of ophthalmologic origin is most frequently an isolated sign, the combination of upper eyelid ptosis (blepharoptosis) and brow ptosis as seen on the painting makes another underlying ophthalmological cause unlikely. Examples of ophthalmological causes of acquired blepharoptosis other than NF1-associated mechanical ptosis include aponeurotic ptosis and traumatic ptosis. Aponeurotic ptosis, also known as senile or age-related ptosis, is the most common cause of blepharoptosis and is caused by the stretching of the levator muscle and its aponeurosis, resulting in a loss of muscle tone and the inability to hold the upper eyelid in the normal position. Traumatic ptosis may be caused by levator disinsertion subsequent to oedema or haemorrhage (mild trauma), the severing of the levator tendon leading to scarring and secondary mechanical ptosis (moderate trauma) , and by damage to the nerve supply of the levator muscle (severe trauma).[9]

However, in addition to the obvious blepharoptosis, the portrait shows an eyebrow region which seems to be pushed slightly downwards, possibly suggesting an underlying frontal sinus pathology.

The Portrait of a One-eyed Man: alternative rhinological diagnoses

From an otorhinolaryngological point of view, several alternative underlying diagnoses are possible which may account for the external deformity of the "one-eyed man". It is most likely that Van Gogh named his painting after a popular nickname that was used in Saint-Remy to refer to this fellow-patient. This assumes that the underlying pathology of the "one-eyed man" was developing slowly, ruling out infectious causes of frontal swelling and ptosis, such as frontal or frontoethmoidal sinusitis with preseptal cellulitis and/or intraorbital complications such as abscess formation in the orbit. Given the assumption of the slow development of the disease and the apparently healthy appearance of the "patient", who even has a visible blush on his right cheek, malignancies of the paranasal sinuses such as adenocarcinoma of the ethmoid with intraorbital and intracranial extension, and a resulting oculomotor nerve paralysis accounting for the ptosis we see, are also less likely.

Given these considerations, the underlying disease resulting in a unilateral eyelid ptosis and alleged frontal swelling in Van Gogh's fellow-patient, is most likely to be of a benign and slowly developing nature, originating from the frontal sinus and possibly extending into the orbit. Three pathologies which can meet these criteria are: a pneumosinus dilatans frontalis, a giant frontal sinus osteoma and a giant frontal sinus mucocoele.

Pneumosinus dilatans is a rare craniofacial malformation which is characterised by the abnormal expansion of the sinus beyond the normal limits of the sinus cavity bone. The frontal sinus is affected most often. However, the involvement of other paranasal sinuses is also possible. Different hypotheses have been proposed to explain the pathogenesis, including the presence of a one-way valve at the sinus entry, local growth disturbances, spontaneous mucocoele drainage, hormonal dysregulation, osteoclastic and osteoblastic activity and trauma.[10] Typical symptoms and signs are frontal bossing and prominence of the supra-orbital ridge accompanying the outward expansion of the pneumosinus. The expansion may also be directed intracranially or towards the orbit, causing headache, diplopia or local pressure symptoms.[11] However, in a review of 17 cases of pneumosinus dilatans involving the frontal sinus, unilateral or bilateral frontal bossing was the most frequent symptom, but none of 17 cases presented with ptosis of the eyelid, making this diagnosis very unlikely.[10]

Osteoma is a benign, slow-growing bone tumour consisting primarily of well differentiated mature, compact, or cancellous bone, most often involving the frontal or ethmoidal sinus.[12] It is the most common benign tumour of the paranasal sinuses with a prevalence of 3%, as observed in CT studies.[13] When larger than 30 mm, these tumours are considered to be giant osteomas. In a review by Cheng *et al*.,[14] 45 patients with giant osteomas were identified in 41 articles, emphasising the rare nature of this pathology. Unlike osteomas of normal size, giant osteomas frequently cause symptoms and even fatal complications. In the review by Cheng *et al*.,[14] 42 cases (93.3%) had serious complications and there was skull involvement (including intracranial mucocoeles, pneumocephalus and cerebral abscess) in 18 cases (40%). There was invasion of the orbit in 24 cases (53.3%). Frontal sinus giant osteomas preponderantly led to intracranial complications (53.6%), while ethmoid giant osteomas often caused intraorbital complications (68.9%). Consequently, ocular signs were very common in patients with giant osteomas of the ethmoid sinuses, including proptosis (40% of total group), diplopia (17.8% of total group), epiphora and eyelid ptosis (8.9% of total group). However, the relatively rareness of eyelid ptosis described in the literature makes the possibility of an underlying giant osteoma less likely, especially when other, more frequent ocular signs such as proptosis are apparently not present (or not visible) on the portrait.

This makes a giant frontal sinus mucocoele a more plausible explanation for the patient's condition. Paranasal sinus mucocoeles are benign, slowly-expanding epithelial-lined lesions containing mucus. These lesions were called mucocoeles for the first time by Rollet in 1896, seven years after Van Gogh painted the portrait of the one-eyed man. Mucocoeles can erode through the surrounding bone and spread intraorbitally or intracranially.[15] It is generally believed that mucocoeles are the result of the obstruction of the sinus ostium subsequent to inflammation, fibrosis, previous surgery, a mass lesion or trauma. If we look at the painting, there is a suspicion of a slight bulging at the level of the radix, possibly suggesting a previous frontal trauma that may have led to a mucocoele, which arises

most commonly in the frontal sinus, followed by the ethmoid sinus.[16] The anatomical proximity to the orbit means that paranasal sinus mucocoeles are often associated with ophthalmic symptoms and signs. In the series studied by Sadiq *et al*.,[16] 45 patients were identified with a diagnosis of paranasal sinus mucocoele, with most of the mucocoeles being found in the frontal sinus (70%). The majority of the 45 patients presented with periorbital swelling and tenderness (64.4%). Proptosis was apparent in 13.3% of cases and diplopia was found in 20% of cases. However, the incidence of eyelid ptosis was not examined and the number of mucocoeles spreading beyond the borders of the paranasal sinuses was not mentioned.[16] Lee *et al*.[6] studied a series of 82 cases with extensive paranasal sinus mucocoeles (defined as sinus mucocoeles with intracranial or intraorbital extension). Intraorbital and intracranial extension were found in 67 and 11 patients respectively. Four patients had both intracranial and intraorbital extension. Frontal and ethmoid sinuses were the two most frequently involved sinuses (45.1% and 28% respectively). Intracranial involvement was caused by frontal or sphenoidal mucocoeles only, but intraorbital extension could be caused by mucocoeles originating in all paranasal sinuses. The most commonly reported symptoms were ophthalmologic symptoms (81 of 82 cases, 98.8%). In contrast to the series of Sadiq *et al*.,[16] the percentage of patients presenting with ptosis was reported and amounted to 33% (27 of 82 cases). Periorbital swelling was reported in 24 cases.[17] Alongside ophthalmologic symptoms, neurological symptoms are less common. When a giant frontal sinus mucocoele extends intracranially by eroding the posterior frontal sinus wall, meningitis, meningoencephalitis, pneumocephalus, brain abcess, seizures, CSF fistula or subdural empyema may be present.[15] Occasionally, a giant frontal sinus mucocoele can manifest with a frontal lobe syndrome (FLS).[18] Frontal lobe syndrome, first defined in 1868 by Harlow, is a pattern of signs and symptoms associated with damage to the frontal lobe. It typically includes a general impairment of planning functions, impulsiveness, lack of inhibition, anti-social behaviour, depression, apathy and perseveration.[18] In short, at the end of the nineteenth century, these symptoms were very likely to be attributed to "mental illness", leading to committal to a mental hospital like the one in Saint-Rémy. Figure 2 shows a frontal sinus mucocoele with intracranial and intraorbital extension.

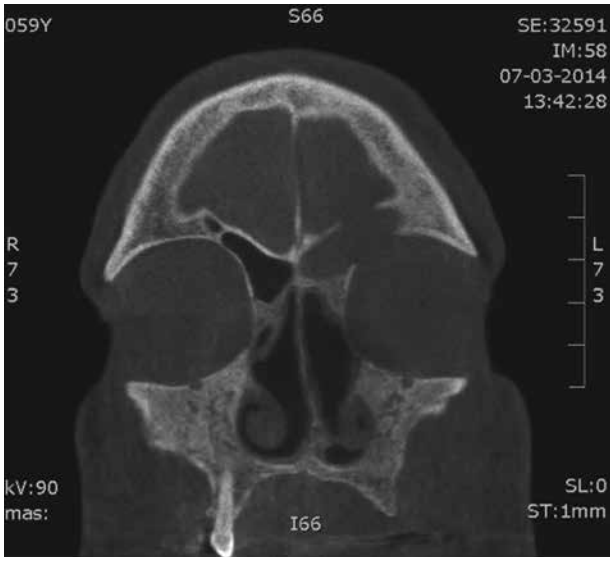

Figure 2
Left-sided frontal sinus mucocoele with limited intraorbital and intracranial extension in a 59-year-old female patient presenting with headache as a primary symptom (cone beam CT of the sinuses, coronal view).

Treatment of frontal sinus pathology at the end of the nineteenth century

If we assume that the "one-eyed man" indeed suffered from a giant frontal sinus mucocoele with intra-orbital extension (leading to ptosis) and intracranial involvement (leading to FMS), how would he have been treated? Five years before Van Gogh painted this portrait, Alexander Ogston described a trephination procedure through the anterior table of the skull to evacuate the frontal sinus, followed by dilation of the nasal frontal duct and curettage of the frontal sinus mucosa (1884). Drainage was established with a tube that was placed in the duct. This technique, which is most suitable for patients with complicated frontal sinusitis, became known as the Ogston-Luc procedure but never gained popularity. However, the presence of a giant frontal mucocoele should have been addressed with a more radical procedure, which was developed in 1895 by Kuhnt, who described the removal of the anterior wall of the sinus in an attempt to clear the frontal sinus disease. In 1898, Riedel and Schenke described the first procedure for the obliteration of the frontal sinus, advocating the complete removal of the anterior table, as well

as the floor of the frontal sinus with stripping of the mucosa. This technique, which led to unsightly cosmetic forehead deformity, was modified by Killian in 1903, who recommended preserving a one-centimeter bar of the supraorbital rim to minimise the cosmetic deformity.[19] However, even if this technique had been available for the one-eyed man, he probably would have died because of the likelihood of severe postoperative complications (CSF fistula, infectious meningitis, meningoencephalitis, brain abscess …), which is probable in cases with intracranial extension of the disease. Nowadays, the majority of patients with giant frontal mucocoeles can be treated safely with minimal complications by means of endoscopic sinus surgery (FESS), including intranasal drainage with marsupialisation and a modified Lothrop (DRAF III) procedure.[17,20]

Conclusion

Because we only have a painted picture of a patient at our disposal, with no history and no possibility of performing a clinical examination or supplemental imaging, our diagnosis can only be considered speculative since the actual underlying pathology will never be confirmed. However, the one thing that is certain and definitive is the splendour of Van Gogh's portrait, which will continue to intrigue all those who see it.

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