SPECIAL ARTICLE

Pediatric Dermatology Vol. 23 No. 3 294–297, 2006

Genuine Diffuse Phlebectasia of Bockenheimer: Dissection of an Eponym

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The scholarly surgeon, Mark Ravitch wrote: "Eponyms are the bane of medical students, the pride of collectors of curiosae and insignificant minutiae, a frequent glib verbal screen for superficial knowledge – or ignorance – a casual name-dropping technique of simulated comprehension, yet withal a guide to the development of medicine, a register of history" (1).

Dr Ravitch was referring to just such an eponym as Bockenheimer syndrome or "genuine diffuse phlebectasia." This term is probably only recognized by a few cognoscenti in the field of vascular anomalies. But as Ravitch admonished, even an expert can be wrong when trying to impress another with an eponym.

Who was Bockenheimer, what condition did he really describe, and does he deserve eponymous status?

ORIGINAL DESCRIPTION

It is unlikely that Philipp Bockenheimer (1875–1933) had eponymic aspirations when he wrote a 1907 report that describes a 52-year-old, barrel-organ player who, since the age of 20, had noted progressive prominence of the veins of the left upper limb (2). The arm had grown to twice the size of the opposite extremity, both in girth and length. The overlying skin was atrophic and the veins had progressively enlarged from the hand to the shoulder. The cephalic vein was prominently dilated (Fig. 1).

Motion of the fingers was limited, yet range of movement in the large joints was normal. The patient complained of incapacitating paresthesias and cramps. Because of ulceration and increasing pain in the palm, resection of a venous mass was performed, along with the radial artery. Postoperatively, the wound failed to heal. Swelling increased and 3 months later the limb was amputated at the glenohumeral joint. The patient died 1 month later, likely related to infection and repeated bleeding.

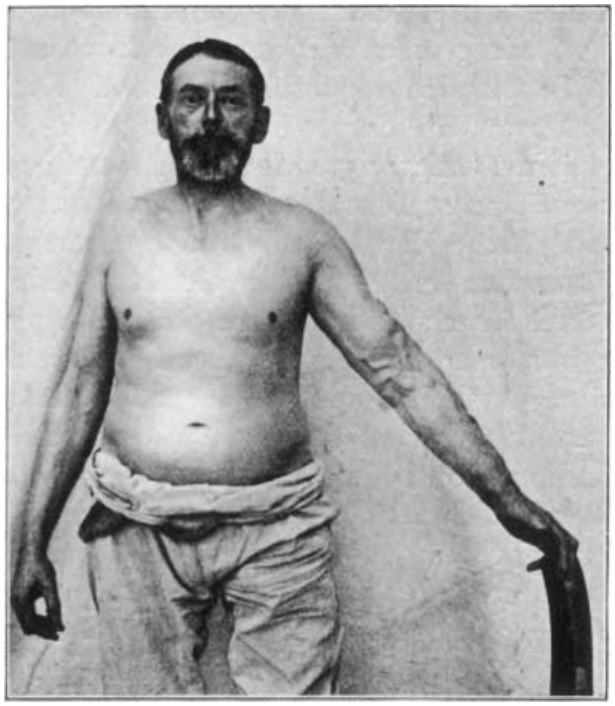

Figure 1. Bockenheimer's patient, reprinted from the original article (2).

Bockenheimer dissected the limb and noted ectasia of all the veins; however, the arteries were normal. The bones were also involved; he observed small holes and

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Figure 2. Dissection of the left arm, colored lithograph, reprinted from original article (2).

Figure 3. Prof. Bockenheimer, Royal Surgical Clinic, Berlin, ca age 40. Prominent scars on the left face probably incurred during initiation to a fencing fraternity. (With permission of the UB der HU zu, Berlin.)

signs of impression by the venous masses (2). The left humerus was 1 cm wider and 3 cm longer than the contralateral side (Fig. 2). Bockenheimer believed this condition was congenital and introduced the designation "genuine diffuse phlebectasia" to differentiate it from late-appearing "venectasias," such as "venous hemangioma" and "varicose veins." The term "genuine" denoted a birth defect in Germanic medical literature of the nineteenth century (3,4).

WHO WAS PHILIPP BOCKENHEIMER?

Philipp Bockenheimer was born on May 26, 1875 in Frankfurt, Germany. He graduated from Berlin Medical School in 1897, and served as assistant at the Robert Koch Institute in Berlin for 2 years. Bockenheimer trained at the Royal Surgical Clinic in Berlin for 8 years before beginning his practice there. He served as surgeon major during World War I and returned to practice at the Royal Surgical Clinic (Fig. 3). He published four papers in the surgical field and wrote three textbooks dealing with standard operative procedures in general surgery (5). However, his passion was anthropology. Despite his busy practice, he found time to take postgraduate courses at the Royal University of Berlin. He was awarded a Doctor of Philosophy in anthropology in 1921. He wrote review papers on anthropology and edited almanacs on South America and Asia (6,7). He lived in Mexico for 10 years, studying anthropology. He was elected a member of the scientific academy Sociedad Academia Nacional de Ciencias Antonio Alzate in 1931, and was awarded a knighthood of Columbus-Orden (Knights of Columbus). Bockenheimer returned to Germany in 1933 and, despite his long

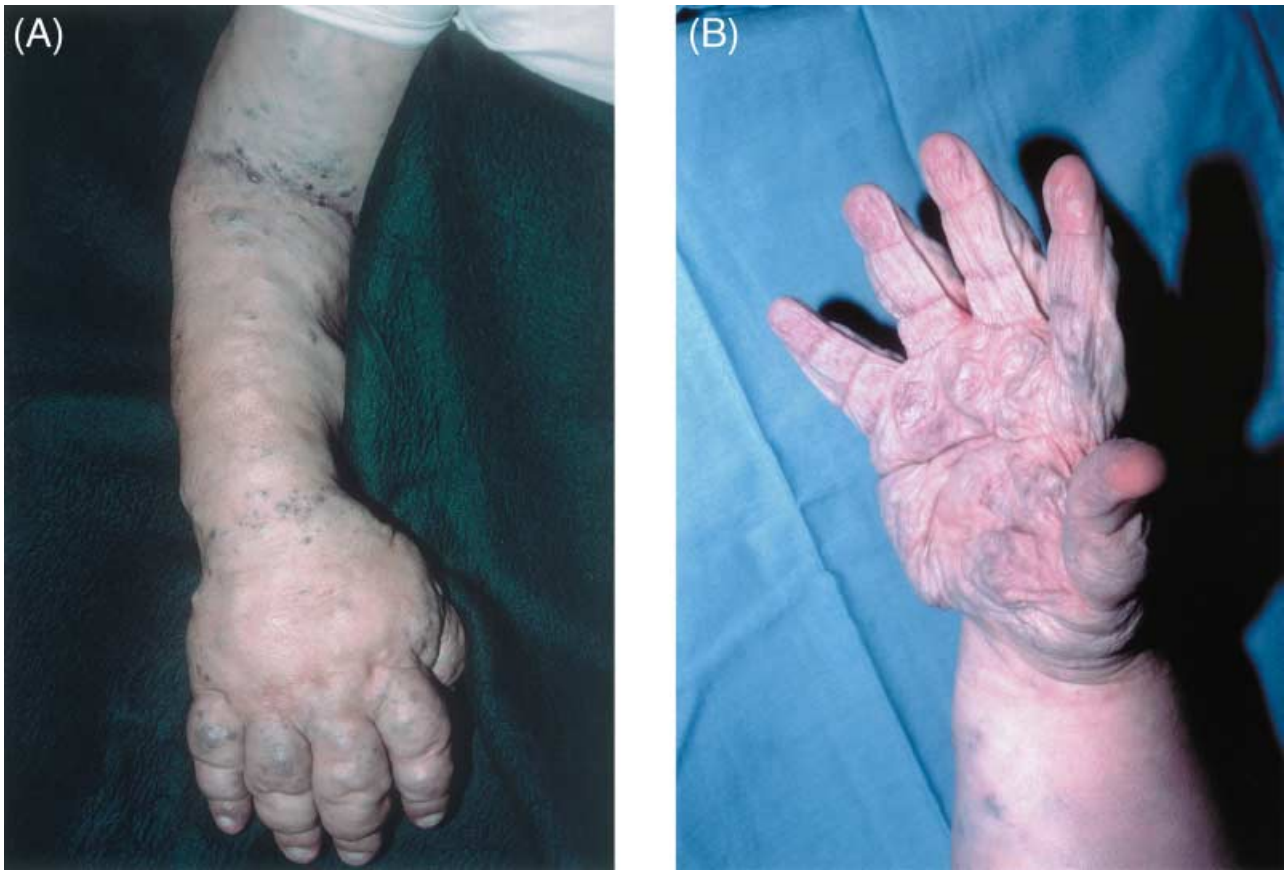

Figure 4. Extensive venous malformation involving all tissues in the upper limb. Note expansion when limb is dependent **(A)** and deflation when arm is elevated **(B)**.

absence, was appointed chief at the "Heiligen Geist Krankenhaus" (Holy Ghost Hospital) in Berlin. He died following a heart attack at age 58 years in Frankfurt on August 12, 1933, leaving behind his wife (8). They had no children.

ASCENSION TO EPONYMOUS STATUS

His colleague Sonntag was first to use the term "Bockenheimer's genuine diffuse phlebectasia" or "Bockenheimer's disease" in 1919 (9). However, as for most eponyms, there are earlier accounts of the same condition. In 1869, Von Pitha (10) described "venous (cavernous) angiectasia" – an extensive vascular anomaly of the extremity with atrophy of muscles and bones – and also mentioned phleboliths as a common finding. In 1889, Palmgren [quoted by Freund (12)] mentioned two cases in his dissertation on "phlebectasia of the upper extremity" that were both "genuine" and "diffuse." In 1906, Heide (11) described a similar patient with "cavernous angioma" of the lower extremity *(*elephantiasis telangiectodes*)*. In 1936, Freund (12) reported an 8-year-old girl with a progressive enlargement of abnormal venous channels in the right shoulder, but the arteries were normal. He noted the unilateral involvement, the relatively frequent association with cutaneous stains, and typical onset in early childhood. Freund concluded that the condition arose as a faulty anlage of an extensive region of the venous vascular system (12). Freund added his patient to the literature and collected 15 other examples: nine in the upper extremity and six in the lower extremity. He also listed clinical criteria for the diagnosis of "genuine diffuse phlebectasia" (of Bockenheimer):

1 Upper limb is more commonly involved than the lower limb.
2 All veins are affected, including the smallest ramifications and without preference for any specific anatomical distribution.
3 Ectasias may be saccular or tubular.
4 Venous dilatation is first noted in childhood and progresses slowly.
5 Histologic examination shows malformed veins with very little muscle and elastin in the media.

6 Thrombosis and phleboliths are common.
7 Affected limb may be long or short; often with wasting of muscle.
8 Pain, swelling, and ulceration can occur.
9 Often there is an associated cutaneous "hemangioma" (sic).
10 Excision of ectatic veins neither helps symptoms nor halts the progression of the disease.

Malan (13) categorized diffuse phlebectasia into two main types: (a) regional and (b) genuine diffuse phlebectasia of Bockenheimer. He underscored that the term "genuine diffuse phlebectasia" denotes that all branches of the venous system in an extremity are affected, including the main deep collecting channels. Mulliken and Young (14) defined genuine diffuse phlebectasia (of Bockenheimer) as "enlargements of all the veins, both large and small, superficial and deep, of part of the body (usually a limb)." In 1990 Roy (15) described patients with "genuine diffuse phlebectasia" of the upper extremity, and classified them clinically and radiographically into three types: (a) circumscribed phlebectasia, (b) regional phlebectasia, and (c) diffuse phlebectasia. He also morphologically separated the phlebectasias into tubular, fusiform, saccular, and plexiform types.

Recent references to Bockenheimer have strayed from his initial description. Enjolras and Garzon (16) referred to "diffuse congenital genuine phlebectasia of Bockenheimer" as a rare disorder in which there are dilated linear veins visible beneath the skin that increase with age. As an example, they illustrated a child with "Bockenheimer syndrome" who had an extensive network of dilated veins over the anterior thorax. Other authors have included "genuine diffuse phlebectasia of Bockenheimer" in the differential diagnosis of cutis marmorata telangiectatica congenita (17–20).

CONCLUSION

"Genuine diffuse phlebectasia of Bockenheimer" and "extensive venous malformation" are synonymous terms that apply to a slow-flow vascular anomaly affecting all tissues in a limb (Fig. 4). The reader can decide whether or not to dazzle a colleague or student with this eponym – whenever the occasion presents.

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