Incidental finding of bifid cardiac apex in a case of sudden death due to sickle cell disease

Kleio Fragkouli1, Antigoni Mitselou1, Vassiliki Boumba1, Theodore Vougiouklakis1,*

Abstract: Bifid cardiac apex represents an infrequent anomaly within human anatomy. When encountered in clinical or forensic practice it has been commonly associated to other cardiovascular malformations. We report an autopsy case of bifid cardiac apex, which was an incidental discovery, in a 29-year-old African male who died suddenly. After histopathological examination death was ascribed to sickle cell crisis. A coronary anatomic variant was also noted. This case is highlighted as a rare representation of bifid cardiac apex and a review of the relevant literature is also presented.

Key Words: bifid cardiac apex, heart anomaly, sickle cell crisis, autopsy.

INTRODUCTION

Normal heart presents ample variations, making it distinctive for each human being [1]. Bifid cardiac apex is an uncommon malformation in otherwise normal human hearts [2, 3]. The first nomenclature of this morphological anomaly dates back in 1875 and was introduced by Rokitansky, as 'apex cordis bifidus' [4].

This entity is encountered in the normal hearts of dugongs, whales, manatees and other marine mammals, possibly as a result of the heart’s adaptation to diving habits [3, 5, 6]. The presence of bifid cardiac apex in humans has been frequently associated with other congenital cardiovascular defects [2, 3, 7-10].

We report an autopsy case of bifid cardiac apex, coexistent with a coronary anatomic variant, found incidentally in an individual who had a sudden death due to sickle cell crisis, without any other major morphological cardiac anomalies. Also, we present a review of the literature which is relevant to the anatomy, occurrence and relationships involving bifid cardiac apex.

CASE REPORT

The deceased was a well-built 29-year-old African male. The body was presented with a history of sudden collapse while hiking; he died on the spot. No other relevant information concerning his past illness and family history was available. On external examination, multiple shallow, rounded or elliptic scars (of maximum diameter 1.5 cm) were observed on the skin over dorsum of both legs and on the medial surface of his wrists (Fig. 1A). Cyanosis of finger nails was present. No other significant external findings were observed. On internal examination, all organs, apart from the heart, showed no major morphological abnormalities. Spleen, except for congestion, displayed no other macroscopic oddities.

His heart weighed 310 g. There was a bifid cardiac apex with a cleft of 1.5 cm length, separating the left and right ventricles (Fig. 2). The ventricles were separated by adipose tissue. All four cardiac chambers were not dilated. Interventricular septa showed no defects. The fossa ovalis was closed. Cardiac valves were unremarkable. Coronary ostia and coronary sinus were normally positioned. The coronary circulation was right-
dominant. The distal part of left anterior descending branch kept its path over the left ventricle, ending at the rim of the cleft. The left coronary trunk measured 25 mm. The left circumflex artery showed an acute take-off from the left coronary trunk.

Toxicological analysis was negative for alcohol and substances of abuse.

All organs were kept for microscopic examination as internal gross findings were inconclusive. Histopathology report revealed sickled red blood cells occluding small-sized blood vessels and arterioles of brain, lungs, liver and kidneys. Additional findings were cerebral edema and lung edema with focal hemorrhage in the interstitium (Fig. 1B). Spleen showed marked congestion of the red pulp and extravasation with pooling of sickled erythrocytes and scattered small aggregates of normocytes, and slight hyperplasia of white pulp without germinal centers (Fig. 1C). Considering the histopathological findings, death was ascribed to sickle cell crisis.

DISCUSSION

Bifid cardiac apex is an infrequent congenital anomaly in human hearts. Human embryology describes the presence of a cardiac notch, which is visible during 5th to 8th intrauterine period, designates the location of the developing interventricular septum and typically disappears by the 11th week of gestation. Its persistence postnatally suggests unusual heart anomalies, such as the bifid cardiac apex [2, 7, 11].

In a search of the English-language literature, we found only a handful of anatomical descriptions in books dating from 1839 to 1912 [4, 6, 11, 12] and few case-reports from 1912 up to nowadays [2, 3, 7-10, 13] describing the bifid cardiac apex. The first reference to this cardiac malformation was noted in 1839 by Reid who reported [6]. “The two grooves (anterior and posterior longitudinal sulci) are connected with each other at or near the apex by a small notch, which is sometimes of sufficient depth to give the heart a bifid appearance.” In 1875, Rokitansky introduces the term ‘apex cordis bifidus’ and designates this anomaly as “a deviation from the normal type in the external form of the heart” resulting from an arrest at an early stage of embryonic development [4]. In terms of prevalence, in 1878 Orth stated that a deep cleft of the cardiac apex is of quite common occurrence and also maintained that “the formation of the apex affords a good indication of the presence or absence of enlargement of the right side of the heart” [12]. Similarly, Mall suggested that “a slight cleft of the apex is not rare, for it is produced whenever the anterior and posterior longitudinal sulci (of the heart) meet” [11]. However, the first documented observation of a bifid cardiac apex was reported by Mall in 1912, who briefly described an adult heart with bifurcation of the apex, presenting a 2 cm-long cleft between the ventricles [11].
Although these former authors support that the bifid cardiac apex is a relatively common malformation, only 9 recorded cases have since surfaced in the literature [2,3,7-10,13]. Among these cases, one is a 27-week-old foetus [7], two are newborns [2], two are children [3,8], and only four are adults [2,9-11,13]. Our case is a rare case of a bifid cardiac apex encountered in an adult.

In the majority of the case-reports previously published (8 out of 9 cases), 'apex cordis bifidus' was combined with major cardiac malformations [2,3,7-10,13]. The most common concomitant abnormalities were atrial and/or ventricular septal defects of various dimensions, observed in individuals of both sexes and aged from newborns to 34 years old [2,3,8-10]. Bifid cardiac apex also has been associated with congenital diaphragmatic hernia in children [14]. The only case where the bifid apex was an incidental finding at autopsy (with no other deformities present) was that of a 25-year-old African-American male who had died from drug intoxication [13].

In the case described herein, the 29-year-old man was eventually found to have died due to sickle cell crisis, a relatively rare cause of sudden death [15]. The precipitating factor for his crisis was probably physical exertion leading to hypoxia, via vascular occlusion, as also described is previous studies [16,17]. The only finding possibly related to sickle cell disease was the skin scars on the legs and wrists but, in the absence of any other relevant macroscopic features, it was considered inconclusive before histopathological examination was performed. Our case presented a bifid cardiac apex at autopsy, with concurrent acute takeoff of the left circumflex artery. Whether a bifid cardiac apex and coronary variants may present as a combination or as an anatomic coincidence remains uncertain. Similarly, whether or not the concurrence of bifid cardiac apex and sickle cell anemia is accidental, poses an interesting query. Further areas of research could shed light into such anatomical or genetic relationships.

In conclusion, bifid cardiac apex is an uncommon finding within human heart anatomy. Our findings involve a bifid cardiac apex concurrent with a coronary anatomic variant in a young male who died due to sickle cell crisis. Bifid cardiac apex may represent an isolated heart anomaly, but awareness of this malformation may also help to seek for other comorbidities from a clinical standpoint.

Conflict of interest. The authors declare that there is no conflict of interest.

References