Pulmonary artery aneurysms in Behçet's syndrome: a review of the literature with emphasis on geographical differences

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ABSTRACT

Objective. *To investigate the frequency of Behçet's syndrome (BS) with pulmonary artery aneurysms (PAA) publications, the most lethal complication of BS, as reported from different countries and to provide a review of diagnostic techniques, treatment approaches and prognosis.*

Methods. *Countries from each continent with a population of 4 million and over were chosen (n=128). A PubMed search for "BS, PAA and the country name" was conducted and 23 countries with BS and PAA were identified. The full texts of articles (n=91) were analysed for data including gender, age, accompanying vascular findings, diagnostic techniques, treatment modalities and mortality rates.*

Results. *A total of 207 (183 males, 24 females) patients with BS and PAA were reported in 91 articles originating from 23 countries. As expected there was a significant correlation (r=0.88, p<0.001) between the total number of articles about BS (n=4431) and those related to PAA and BS. In a simple linear regression analysis the number of BS and PAA articles from Japan was significantly below the identity line while in Turkey there was a propensity to publish more articles related to PAA than expected. One hundred and sixteen patients (56%) were treated with immunosuppressive therapy. Biologics were used only in 5 patients (2%). Of the 207 patients, 62 (30%) died.*

Conclusion. *PAA is mostly reported as case reports from countries where BS is common. PAA might be uncommon in Japan. The prognosis of PAA could be getting better.*

Introduction

Behçet's syndrome (BS) is most frequently seen in the Mediterranean, Middle East, and the Far East. The highest prevalence rate was reported from Turkey as 80–370 per 100,000 population (1). The prevalence ranges from 2–30 cases per 100,000 in other Asian countries, with lower figures in Europe and the USA (2).

BS can have diverse clinical manifestations at differing geographies. O'Neill *et al*. suggested that BS patients from Middle Eastern countries had lower rates of arthritis, vascular problems, and central nervous system abnormalities compared to patients from Western countries (the UK and USA) (3). In another study focusing on geographic differences, eye and vascular disease, however, were similar in frequency in both USA and Turkish patients (4). Krause *et al*. showed that patients from North Africa had higher rates of arthritis, overall vascular disease, deep vein thrombosis, and neuro-Behçet in contrast to patients with origins in Iran, Iraq and Turkey (5). It also has been reported that in Japan the frequency of vascular involvement in BS is low (6).

Pulmonary artery aneurysm (PAA) is a rare vascular complication of BS. In previous studies reported from Turkey, the prevalence rate of PAA in BS was observe to between 0.16% and 1.1% (7-8). PAA, the most lethal form of disease expression in BS, is not usually investigated in a patient with BS unless the patient has haemoptysis or thrombotic events. It follows that a clinician needs at least an approximate estimate of the frequency of this potentially lethal complication in the setting where he/she practices. Unfortunately, such data are not available.

With the current work we sought to seek this information in an indirect fashion. We hypothesised that in the geographies where BS is more common, there would be more medical publications about associated PAA. Thus the first aim of this study was to investigate the frequency of the cases of BS with PAA as it was reported from different countries.

The second aim, about which there has also been few formal data from differ-

ing geographies, was to evaluate the diagnostic modalities, and treatment approaches used, along with prognosis by literature review.

Methods

Countries from each continent with a population over 4 million were chosen (n=128). A PubMed search for "BS, PAA and the country name" was conducted and 23 countries with BS and PAA were identified. In the initial step, 126 studies were identified and 35 of these were excluded after reading their abstracts. Among those 35 studies, 5 were eliminated because the arterial aneurysms were at sites other than the pulmonary arteries and 13 were excluded because they were review articles. An additional study was left out since it was a cohort study about the general characteristics of vascular involvement in BS. One study was not included because it was a commentary only. In two studies which were written in languages other than English, the abstracts did not give enough information. Finally, 13 studies which were not directly about cases of BS associated with PAA were left out.

To address the second aim of our study, full texts of the remaining articles (n=91) related to BS with PAA were analysed for demographic data including gender, age, ethnicity in addition to accompanying vascular findings, diagnostic modalities and treatment. The reported mortality rates were also tabulated. Articles about 5 patients or less were listed as case reports while the rest made up the case series.

Results

There were a total of 207 BS cases (183 males and 24 females) reported with PAA (mean age 31.7±9.3 (SD)). These were presented in 98 case reports and in 8 case series. The studies with case series were published only from Turkey (n=7) and France (n=1). The number of cases in these articles were 26, 24, 15, 13, 13, 8 and 5 for the former and 5 for the latter. Most of the cases were also reported from Turkey (n=135) (65%). The ethnic origins of the patients from each country are also shown in Table I. Ninety-one studies related to BS with PAA were published from 23 countries between 1981 and 2014 with the first reported case of BS with PAA being from France. When we looked at a simple linear regression analysis, there was a significant correlation between the numbers of the articles for BS (n=4431) and BS associated with PAA for each country (r=0. 88, $p<0.001$). Figure 1 shows the regression line with the 95% confidence intervals. It is to be noted that there was a rather good linear correlation between the total number of articles and those related to PAA. The 2 countries particularly away from the

Table I. Number of cases with BS and PAA from each country and demographic features of patients.

Continent	Country	Year	P(n)	CR(n)	CS (n)	TC (n)	Gender	Mean age	Ethnicity
ASIA	China	2014	1	1	0	1	M	28	NG
	India	2002	1	1	0	1	M	28	NG
	Japan	1990-2009	5	5	0	5	5 M	43 ± 13	1 Japanese, 4 NG
	Turkey	1993-2010	29	31	7	135	123 M, 12 F	33 ± 6	NG
	Iran	2007-2013	4	4	0	4	3 M, 1 F	38 ± 6	4 NG
	Israel	1988-2009	4	4	0	4	3 M, 1 F	18 ± 5	2 Arab, 2 NG
	Saudi Arabia	2009-2010	2	2	0	2	1 M, 1 F	32 ± 9	2 Arab
	Korea	1994-2014	6	6	0	6	3 M, 3 F	33 ± 12	2 Korean, 4 NG
AFRICA	Egypt	2007-2013	2	3	0	3	3 M	21 ± 4	3 NG
	Morocco	2002-2012	6	7	0	7	7 M	30 ± 14	2 Moroccan, 5 NG
	Tunisia	1991-2013	4	4	0	4	4 M	27 ± 6	3 Tunisian, 1 NG
EUROPE	Germany	2003	1	1	0	1	M	26	NG
	France	1981-2004	7	10	1	15	14 M, 1 F	27 ± 7	2 Algerian, 2 French, 5 Moroccan, 1 Turkish, 1 Tunisian, 4 NG
	UK	2005-2011	3	3	0	3	2 M, 1 F	36 ± 6	1 Asian, 2 White
	Italy	2013	1	1	0	1	M	37	NG
	Spain	2011	1	1	0	1	F	29	NG
	Switzerland	1989-2006	2	2	0	1	1 M, 1 F	31 ± 6	Austrian
	Belgium	1998	1	1	0	1	F	35	Caucasian
	Ireland	2004	1	1	0	1	M	48	Irish
NORTH AMERICA	USA	1998-2013	7	7	0	7	6 M, 1 F	26 ± 10	2 African-American, 1 Iranian, 1 Algerian, 1 Moroccan, 1 Chinese, 1 NG
SOUTH AMERICA	Brazil	1988	1	1	0	1	M	25	Brazilian
	Colombia	2010	1	1	0	1	M	18	NG
OCEANIA	Australia	2010	1	1	0	1	M	60	NG
TOTAL	1981-2014	91	98	8	207	183 M, 24 F	32 ± 9		

P: publication; CR: case report; CS: case series; TC: total case; NG: Not given.

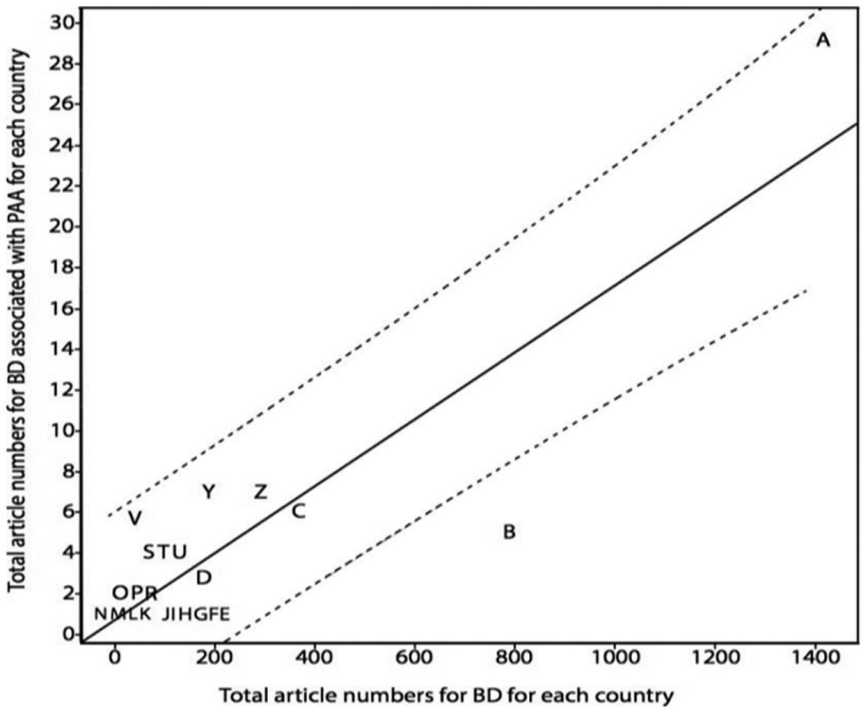

Fig. 1. The distribution of the countries according to the numbers of article published for BS *versus* BS associated PAA from 23 countries (95% confidence intervals).
A: Turkey, B: Japan, C: Korea, D: UK, E: Italy, F: China, G: Germany, H: Spain, I: India, J: Brazil, K: Australia, L: Belgium, M: Ireland, N: Colombia, O: Switzerland, P: Egypt, R: Saudi Arabia, S: Tunisia, T: Iran, U: Israel, V: Morocco, Y: France, Z: USA.

identity line are Turkey and Japan with the former showing a propensity to publish more BS articles related to PAA than expected from the total number of articles related to BS, in general. On the other hand, the number of PAA as a complication of BS articles from Japan are significantly below the identity line (outside the 95% confidence intervals) indicating that the PAA complication in Japanese patients with BS is less than expected as judged by the frequency of BD patients otherwise reported.

In 109 out of the 207 cases (53%), information about the disease duration for BS was available and the mean disease duration of the patients up until the onset of pulmonary manifestation was 5.0±3.0 (SD) years. The mean interval time between diagnosis of BS and the onset of the manifestations of PAA was 3.5±3.0 (SD) years in 89 out of the 207 cases in which the information was provided (43%). Among those 89 cases in which the interval time was given, 12 cases (13%) presented with PAA symptoms as the initial complaint.

In 10 of the 207 cases (5%), patients were 16 or younger, 4 were female. Three out of 4 patients who were reported from Israel were in this pediatric group. Among the remaining patients, 2 cases were from Turkey, 2 were from the USA, 1 from Morocco, 1 from Egypt and 1 was from France.

Accompanying vascular findings in BS patients with PAA are shown in Table II. One hundred and eighteen patients (57%) had concomitant extra pulmonary venous thrombosis which was the most common accompanying vascular finding. In addition, intracardiac thromboses were found in 20 patients (10%) and this was more common among patients from Morocco. Cerebral vasculitis was reported in 2 patients (1%) from India and Saudi Arabia. Arterial aneurysms at other sites were described in 10 patients (5%). Popliteal artery thrombus was an accompanying lesion in 2 patients (1%) from the USA.

Diagnostic procedures used for the diagnosis of PAA in BS patients are shown in Table III. Computed tomography (CT) was the preferred diagnostic technique used. While pulmonary angiography (PA) was mostly used in patients from France, in 60% of the patients (9 out of 15), digital subtraction angiography (DSA) and MR angiography (MRA) were more commonly used in patients from Turkey, in 14% of the patients (19 out of 135) and in 16% of the patients (21 out of 135) respectively. Open diagnostic lung biopsy was reported in 2 (1%) patients from Israel.

Treatment modalities are shown in Table IV. One hundred and sixteen patients (56%) were treated with only immunosuppressive therapy. In 97 out of the 116 patients (84%), cyclophosphamide alone or along with corticosteroids was used. Biologics were used only in 5 patients (2%), from Korea, Spain, Switzerland, UK and the USA. One patient received adalimumab while the remaining 4 were treated with infliximab. Additionally, in 20 (10%) patients, embolisation was chosen alone or with immunosuppressive therapy and this method was used in patients from France more than from other countries.

Lastly, 46 (22%) patients underwent surgery, which was more common in Iran and Israel. In 18 of those who underwent surgery (39%), immunosuppressive treatment was also used before or after surgery.

Of the 207 patients, 62 (30%) patients died and the death in 17/62 (27%) patients was during or in a few months after surgery as particularly reported from Turkey 11/43 (26%), France 1/6 (17%), Iran 1/2, Israel 2/2, Morocco 1/2 and Tunisia 1/1. In 47 out of the 62 (76%) patients who died and in whom survival data was available, the life span was 9.0±6.0 (SD) months after the onset of lung symptoms.

Discussion

PAA is a rare condition and, as expected, is mostly reported from countries where BS is common. A good number of articles about PAA associated with BS are presented as "case reports". In our survey there were only 8 articles reporting case series and the highest number of patients in any one case series was limited at 26. Turkey had the highest number (65%) of reported cases of PAA in this series. The proposal that the overall vascular complications are at lower rates in the Middle Eastern countries needs further verification (5).

Table II. Accompanying vascular findings of patients with BS and PAA*.

Continent	Country	Case (n)	Accompanying vascular findings	n (%)
ASIA	China	1	Arterial aneurysm	1
	India	1	Cerebral vasculitis	1
	Japan	5	Venous thrombosis	3 (60)
			Intracardiac thrombus	1 (20)
	Turkey	135	Venous thrombosis	86 (64)
			Intracardiac thrombus	6 (4)
			Arterial aneurysm	6 (4)
	Iran	4	Venous thrombosis	1 (25)
			Arterial aneurysm	1 (25)
	Israel	4	Venous thrombosis	1 (25)
			Intracardiac thrombus	2 (50)
	Saudi Arabia	2	Venous thrombosis	1 (50)
			Cerebral vasculitis	1 (50)
	Korea	6	Venous thrombosis	4 (67)
			Intracardiac thrombus	1 (17)
AFRICA	Egypt	3	Venous thrombosis	2 (67)
	Morocco	7	Venous thrombosis	2 (29)
			Intracardiac thrombus	5 (71)
			Arterial aneurysm	1 (14)
	Tunisia	4	Venous thrombosis	3 (75)
			Intracardiac thrombus	2 (50)
EUROPE	Germany	1		0
	France	15	Venous thrombosis	7 (47)
	UK	3	Venous thrombosis	2 (67)
			Arterial aneurysm	1 (33)
	Italy	1		0
	Spain	1		0
	Switzerland	2	Intracardiac thrombus	1 (50)
	Belgium	1	Venous thrombosis	1
	Ireland	1	Venous thrombosis	1
NORTH AMERICA	USA	7	Venous thrombosis	3 (43)
			Intracardiac thrombus	2 (29)
			Popliteal artery thrombus	2 (29)
SOUTH AMERICA	Brazil	1	Venous thrombosis	1
	Colombia	1		0
OCEANIA	Australia	1		0
TOTAL		207	Venous thrombosis	118 (57)
			Intracardiac thrombus	20 (10)
			Arterial aneurysm	10 (5)
			Cerebral vasculitis	2 (1)
			Popliteal artery thrombus	2 (1)

*More than one accompanying vascular findings were in some cases.

Table III. Diagnostic procedures used for the diagnosis of PAA*.

Diagnostic procedures	n (%)
Computed tomography	154 (74)
CT angiography	14 (7)
MR angiography	24 (12)
Digital aubtraction angiography	22 (11)
Pulmonary angiography	35 (17)
Open lung biopsy	2 (1)

*More than one diagnostic procedure was used in some cases.

Our indirect approach to the epidemiology of PAA among different geographies gave interesting results. Our study suggests that the number of publications in PubMed could be useful in having a rough estimate of the prevalence the frequency of a specific feature of a disease. In this line it was particularly interesting to observe that the number of publications related to PAA from Japan was significantly less than what could be estimated from the total number of publications from that country. This is in line with a previous survey of PAA among patients with vascular disease from Japan which reported that PAA was indeed quite rare in this country, a geography well known for its high frequency of BS (6).

Previous studies reported that BS ran a more severe course in young men. Yazici *et al*. showed that male sex and younger age at disease onset (≤25 yr) were associated with more severe disease (9). Dilsen *et al*. described a higher prevalence of vital organ involvement in male patients (10). Furthermore the preponderance of males in BS with PAA also has been emphasised in many reports. The average age at the time of diagnosis of BS with PAA is 30 years (11). In line, in the current survey, 88% of the patients were male and the mean age of onset of PAA of these patients was similar to those previously reported.

An important clinical feature of patients with PAA is the coexistence of other, extra pulmonary large vessel lesions (12). In our review, the association between pulmonary artery aneurysms and venous thrombosis was seen in 57% of the patients whereas a previous paper reported more than 80% of patients with PAA had coexistent venous thrombosis of the lower extremities or vena cavae (13). Moreover, intracardiac thrombus in patients with PAA has also been reported. Cases complicated by intracardiac thrombus have mostly originated from the Mediterranean basin and the Middle East (14). In a previous study, it has been reported that 12% of the patients with PAA had intracardiac thrombus (13). We found similar rates to those reported earlier with 10% of patients having intracardiac thrombus and they were also more commonly from Mediterranean countries.

The diagnostic techniques used for the BS patients with PAA were somewhat different among the countries. The most common modality was CT, used in 74% of all patients. CT is preferred most because of its non-invasive nature, ease of performance and increasing availability. It also demonstrates high quality of vascular images with minimal amount of contrast material used (15). Conventional angiography once regarded as a gold standard for the diagnosis of PAAs was used only in 17% of the patients, most of whom were from France. Angiography is well known to carry the risk of aneurysm rupture (16) and the potential of causing vascular injury through

Table IV. Treatment modalities used for the BS patients with PAA.

Continent	Country	Case (n)	Immsup (n) (%)	Biologic (n) (%)	Embolisation (n) (%)	Surgery (n) (%)	Mortality (n) (%)	Mortality-related surgery (n) (%)
ASIA	China	1	0	0	0	1	0	0
	India	1	1	0	0	0	0	0
	Japan	5	3 (60)	0	0	1 (20)	2 (40)	0
	Turkey	135	90 (67)	0	8 (6)	19 (14)	43 (32)	11/43 (26)
	Iran	4	1 (25)	0	0	3 (75)	2 (50)	1/2
	Israel	4	1 (25)	0	0	3 (75)	2 (50)	2
	Saudi Arabia	2	0	0	1	1	0	0
	Korea	6	2 (33)	1 (17)	0	3 (50)	2 (33)	0
AFRICA	Egypt	3	2 (67)	0	0	1 (33)	0	0
	Morocco	7	4 (57)	0	0	2 (28)	2 (29)	1/2
	Tunisia	4	1 (25)	0	1 (25)	2 (50)	1 (25)	1
EUROPE	Germany	1	0	0	0	1	0	0
	France	15	6 (40)	0	6 (40)	3 (20)	6 (40)	1/6 (17)
	UK	3	1 (33)	1 (33)	0	1 (33)	0	0
	Italy	1	0	0	1	0	0	0
	Spain	1	0	1	0	0	0	0
	Switzerland	2	1	1	0	0	0	0
	Belgium	1	1	0	0	0	0	0
	Ireland	1	0	0	0	1	0	0
NORTH AMERICA	USA	7	2 (29)	1 (14)	2 (29)	2 (29)	2 (29)	0
SOUTH AMERICA	Brazil	1	0	0	0	1	0	0
	Colombia	1	0	0	0	1	0	0
OCEANIA	Australia	1	0	0	1	0	0	0
TOTAL		207	116 (56)	5 (2)	20 (10)	46 (22)	62 (30)	17/62 (27)

the pathergy phenomenon. The reason that the French authors used it frequently was that they also used embolisation rather frequently to manage PAA. The other diagnostic techniques used were MR angiography (MRA) and digital subtraction angiography (DSA) which are noninvasive and more commonly preferred in patients from Turkey.

In our survey, immunosuppression by cytotoxic drugs had been the main modality of treatment, usually in combination with glucocorticoids. Such treatment has the potential to stabilise small aneurysms in the pulmonary vasculature, and in some cases can even make them regress (15). This treatment modality was also the most commonly used regimen in this survey, in 116/207 (56%) of the patients. Among these cyclophosphamide has been the most frequently used (84%) cytotoxic immunosuppressive. It is to be noted that there are no randomised controlled studies to evaluate treatment options in PAA. This combination of cyclophosphamide with glucocorticoids is in line with EULAR recommendations for managing BS (17).

Our survey revealed that biologic treatment was used rather sparingly, only in 5 patients. It is to be noted that all 5 had favourable outcomes.

Surgery as a life-saving procedure when medical therapy fails, carries a high risk of mortality (18) in managing PAA in BS. Twenty seven percent of all mortality (17 out of 62) in this survey followed surgery. In a comprehensive literature search of articles on the cardiothoracic procedures for cardiovascular complications of BS, the postoperative morbidity, recurrence and mortality rates were 21.4%, 11.7% and 15.0% (19). Transcatheter arterial embolisation is a less invasive alternative method to surgery in selected cases of PAA (20). In our survey, embolisation was chosen alone or with immunosuppressive therapy in 10% of the patients. PAA has always been considered to be the most mortal complication of BS. It has been reported that half of the patients died within 10 months of the onset of haemoptysis (13). In another study, the estimated mortality rate was 60% (21). In the current survey, however, we observed a considerably better overall mortality rate of 30% and this probably, indicates a globally increased awareness and hence more prompt management of this lethal complication. On the other hand, similar to previous studies, the life expectancy of PAA patients was only 9 months after the onset of pulmonary aneurysm symptoms.

Our study has some limitations. Since it includes only reported cases any patient with a bad outcome may have been underreported. In addition, the reported cases could have special or uncommon manifestations. Our judgment about the use of surgery should be viewed with caution. The surgery was probably undertaken because of more severe and life-threatening disease in many of these patients.

In conclusion, this study is the first report of a literature review regarding the BS patients with PAA presented from different countries around the world and it supports our notion that what is published can be a clue to epidemiologic features of a disease, also supporting a previous contention that PAA might indeed be uncommon in Japan. It

also suggests that the prognosis of PAA might be getting better with the possibility that a more liberal use of biologics will bring about a better prognosis.

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