Pathophysiology, clinical evaluation and treatment options of spontaneous pneumothorax

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ÖZET

Spontan pnömotoraksın patofizyolojisi, klinik değerlendirmesi ve tedavi seçenekleri

Apikal subplevral bleb primer spontan pnömotoraksın en sık sebebi iken, kronik obstrüktif akciğer hastalığı sekonder spontan pnömotoraksın en sık sebebidir. Tanı fizik muayene, akciğer grafisi ve torakoskopik muayene ile doğrulanır. Tipik olarak astenik ve uzun yapılı kişilerde görülür. Spontan pnömotoraks katameniyal pnömotoraks ve gebelik süreci gibi hayatın özel dönemlerinde görülebilir. Primer spontan pnömotoraksın tedavi seçenekleri konservatif, ara ve cerrahi prosedürleri içermektedir. Sonuç olarak, spontan pnömotoraksın patofizyolojisini, klinik değerlendirmesini ve tedavi seçeneklerini literatür ışığında bir derleme ile sunmayı amaçladık.

Anahtar Kelimeler: *Spontan pnömotoraks, video yardımlı torakoskopik cerrahi, bül, derleme.*

SUMMARY

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An apical subpleural bleb is most common pathology of primary spontaneous pneumothorax however, chronic obstructive pulmonary disease is most common cause of secondary spontaneous pneumothorax. The diagnosis is confirmed by physical examination, chest radiography and thoracoscopic examination. The typical person who present has an asthenic body, being taller and thinner than the average person. Spontaneous pneumothorax can seen in some special part of life such as; catamenial pneumothorax and during pregnancy. Therapeutic options of primary spontaneous pneumothorax is include the conservative, intermediate and invasive procedure. In conclucion we aimed the report pathophysiology, clinical evaluation and treatment options of spontaneous pneumothorax in the light of literatures with a review article.

Key Words: *Spontaneous pneumothorax, video assisted thoracoscopic surgery, bullae, review.*

DEFINITION and PATHOPHYSIOLOGY

Collapse of one or both lungs, caused by accumulation of gas or air in the pleural cavity resulting from injury or diseas is definition of pneumothorax. Spontaneous pneumothorax (SP) classified primary and secondary. Primary spontaneous pneumothorax (PSP) usually occurs with young people who are healthy. Most common pathology is the rupture of an apical subpleural bleb (1). Bronchiolar inflammation, fibrosis and high mechanical stresses at the apex of lung are reasons of the rupture of an apical subpleural bleb (2). Inflammatory changes in the distal airways play an important role in the occurrence of the pneumothorax during transpulmonary pressure changes (3,4). Secondary spontaneous pneumothorax (SSP) is associated with people who have lung disease, the most common cause is chronic obstructive pulmonary disease, metabolical diseases, malignancies and infections diseases (2).

CLINICAL EVALUATION

The incidence of PSP is 7.4/100.000 per year in men and 1.2/100.000 per year in women. The incidence of SSP is 6.3/100.000 per year in men and 2.0/100.000 per year in women (5). Smoking is very important risk factor. The relative risk of pneumothorax is 7 times higher in light smokers (1-12 cigarettes per day), 21 times higher in moderate smokers (13-22 cigarettes per day), 80 times higher in heavy smokers (> 22 cigarettes per day) (2). A positive family history is found in 11.5 % of who present with a SP (2).

The disease is clinically silent, unless one of the blebs ruptures and causes a pneumothorax (2). Many patients do not seek medical advice for several days, 46% waiting more than 2 days before presentation despite symptoms (1). The symtoms are chest pain, coughing, deep breathing, rapid heart rate, and fatigue (6). The chest pain is first and most common symptom, because of the rupture of an apical bleb, causing the air leak, releases irritant material into the pleural cavity, eosinophilic infiltration and stimulating inflammation of the parietal pleura (7). The tension pneumothorax presents with severe sudden beginning, anxiety, swollen neck veins, weak pulse, and decreased breathing sounds from the lung, reversible horners syndrome reported with SP, the symptoms of SSP are more serious than those of PSP because of include the lung disease (6,8,9).

The diagnosis is confirmed by physical examination, chest radiography and thoracoscopic examination (10). Chest radiograph is a standard diagnostic procedure of pneumothorax. An asthenic body, being taller and thinner than the average person with suddenly chest pain and shortness of breath is typical person of SP. Electrocardiographic changes may develops at SP. Left sided pneumothorax include a rightward shift of the frontal QRS axis, reduced precordial R wave amplitude, decreased QRS amplitude and precordial T wave inversion and so it is simulating of a myocardial infarction (11,12).

SPECIAL CLINICAL ENTITY with SPONTANEOUS PNEUMOTHORAX

SP can seen in some special part of life and clinical entity, such as; catamenial pneumothorax, during pregnancy, in Acquired Immune Deficiency Syndrome (AIDS), relationship with aluminum level and meteorological conditions (13-18).

Catamenial Pneumothorax

Catamenial pneumothorax is a specific type of SSP. It is characterized by a temporal relationship with menses. It is most frequent symptom of thoracic endometriosis. It occurs mainly in women above 30 years of age and is typically right sided. During video-assisted thoracoscopic surgery (VATS), inspection of the diaphragmatic surface is paramount, plication of the involved area alone can be successful for treatment. In complicated cases, hormonal suppression therapy can be helpful (13).

Spontaneous Pneumothorax and Pregnancy

Though more common in male patients, PSP might be expected to occur reasonably often in female patients of child bearing age. Special risks are posed for both the mother and the fetus in this situation. Management strategies is very important, when the SP become in pregnancy three important practical issues need to be addressed. How should PSP be managed if it occurs during pregnancy? How should the subsequent delivery be managed? and is a surgical procedure required postpartume? So the surgeon must choose the best management procedures for mothers and infants, observation, simple aspiration, small bore chest drains, elective surgery at or near term with regional anesthesia and VATS can choose for treatment (14).

Spontaneous Pneumothorax in AIDS

SP is a frequent complication in non treated human immunodeficiency virus (HIV) infected patients as a complication of opportunistic infections and tumours. Cigarette smoking, injection drug use, pneumocystis carinii pneumonia, pulmonary tuberculosis, pneumatoceles, or bullae are risk factors for SP. The incidence of SP in AIDS is between 2 and 7 per 1000 person-years. Conservative management is associated with high failure rates and prolonged hospitalization. Needle drainage, tube thoracostomy, heimlich valve, pleurodesis, pleurectomy, VATS and thoracotomy can be used in the treatment of pneumothorax. Recurrence rates of SP in AIDS range between 11 and 65%. The overall mortality rate of patients with pneumothorax in AIDS is about 34% (15).

Aluminum and Spontaneous Pneumothorax

Aluminum is known as a toxic metal. Han et al. has reported the relationship between aluminum and SP. It has already been determined to cause asthma bronchial and chronic obstructive pulmonary disease. It was also reported that Shaver disease undergoing fibrosis with large bubbles and the symptoms of potroom asthma had developed in the workers working in the manufacture of aluminum. The aluminum level in blood plasma in SP was significantly higher than that in the control group ($p< 0.001$). In the SP cases, the specificity of the aluminum was determined to be 100% and its sensitivity was determined to be 90%. The aluminum risk level in SP group was determined to be ten times as high as that in the control group. So aluminum plays a role in SP etiology causing the development of subpleural bleb, bullous lesions and chronic obstructive pulmonary disease (16). On the other hand the other study from Leo et al. has not confirmed the relationship between aluminum and SP (17).

Meteorological Conditions and Spontaneous Pneumothorax

The relationship meteorological conditions and SP was reported for a long time in literature, but in recent years new studies are reported about this issue. SP is generally thought to result from the rupture of blebs or bullas. It has been suggested that the rupture of the alveolar walls in diseased portions of the lung that is characterized by the porosity of visceral pleura may represent another pathogenic mechanism. The factors responsible for the initiation of air leaks are not known, but it is believed that rupture may occur when a substantial transpulmonary pressure gradient is present, the pressure equilibrium between surrounding structures and air trapped in blebs, bullas, or diseased alveoli fails to be reached rapidly, resulting in rupture. Conditions associated with the check-valve phenomenon are known to predispose a patient to pneumothorax. Exposure to rapid changes of environmental pressure (scuba diving or flying) may also result in pneumothorax in healthy individuals. Significant variations in possible impact of changes in

atmospheric pressure may be involved in the pathogenesis of SP. The knowledge of this relationship helps the understanding of the pathogenesis of SP (18).

TREATMENT OPTIONS

British Thoracic Society, American College of Chest Physicians, Belgian Society of Pneumology have reported the recommendations for the management of SP, but these three published guidelines include different recommendations (Table 1) (1). Therapeutic options of PSP is variations, it is include the conservative, intermediate and invasive procedure (19). An algorithm of treatment options of PSP has reported by Baumann and Noppen (20) (Figure 1).

Quantification of Pneumothorax Size

Quantification of pneumothorax size is important for decision the type of managements. The exactly quantification of pneumothorax size is very difficult with chest X-ray. Computed tomography is not routinly but it is too sensitive according to chest X-ray (21). The light index is commonly used in europe. It suggests that the volume of a pneumothorax can be calculated as 100-[(average diameter of lung^3/average diameter of hemithorax^3) x 100] (1). The Rhea method is preffered in USA. It uses the average of the interpleural distances measured in centimetres at the apex, midpoint of the upper half of the lung and midpoint of the lower half of the lung on an erect chest X-ray radiograph to pneumothorax size in percent using a nomogram (1). In recent years a new method for estimating the volume of a pneumothorax have been developed using helical computed tomography, estimate pneumothorax size in %= 4.2 + [4.7 x (interpleural distances in cm at apex, midpoint of upper half of collapsed lung and midpoint of lower half of collapsed lung)] (1,22).

CONSERVATIVE MANAGEMENTS

Observation and Conservative Treatment

It is prefer that 20% or less volume PSP in patients without respiratory components. The rate of resolution/reabsorption of PSP was previously estimated as 1.25-1.80% of the volume of hemithorax every 24 hours. Conservative management include the avoidance of the need for hospitalization and associated cost savings (1,23). The rate of recurrence is 16-52%, most recurrences seeing within six months to two years, the rate of recurrences increases with more episodes (24).

Aspiration

Aspiration of PSP by the placement of a needle or more often a catheter into the pleural space. Successful reexpansion of the lung after simple aspiration is 50-83% (1,25). Successful aspiration depend on age and size of the pneumothorax, age under 50 years 70-81% success, over 50 years 19-31% success and size of the pneumothorax < 3 L aspirated 89% success, > 3 L no success, > 50% size on chest film 62% success, < 50% size on chest film 77% success (1,26). Simple aspiration include some complication such as local subcutaneous emphysema, vasovagal reactions, catheter kinking, dislodgement, empyema and re-expansion pulmonary edema but has an advantage like shorter hospital stay (1).

Table 1. Comparison of guideline recommendations (clinically stable patients) (1).

Guideline	Small primary spontaneous pneumothorax	Large primary spontaneous pneumothorax
British Thoracic Society	Conservative management as outpatient	Simple aspiration
American College of Chest Physicians	Observation in emergency department followed by conservative management as an outpatient	Pleural catheter insertion (small bore or intercostal catheter) and drainage
Belgian Society of Pneumology	Conservative management as outpatient	Aspiration or pleural catheter insertion (small bore or intercostal catheter) and drainage

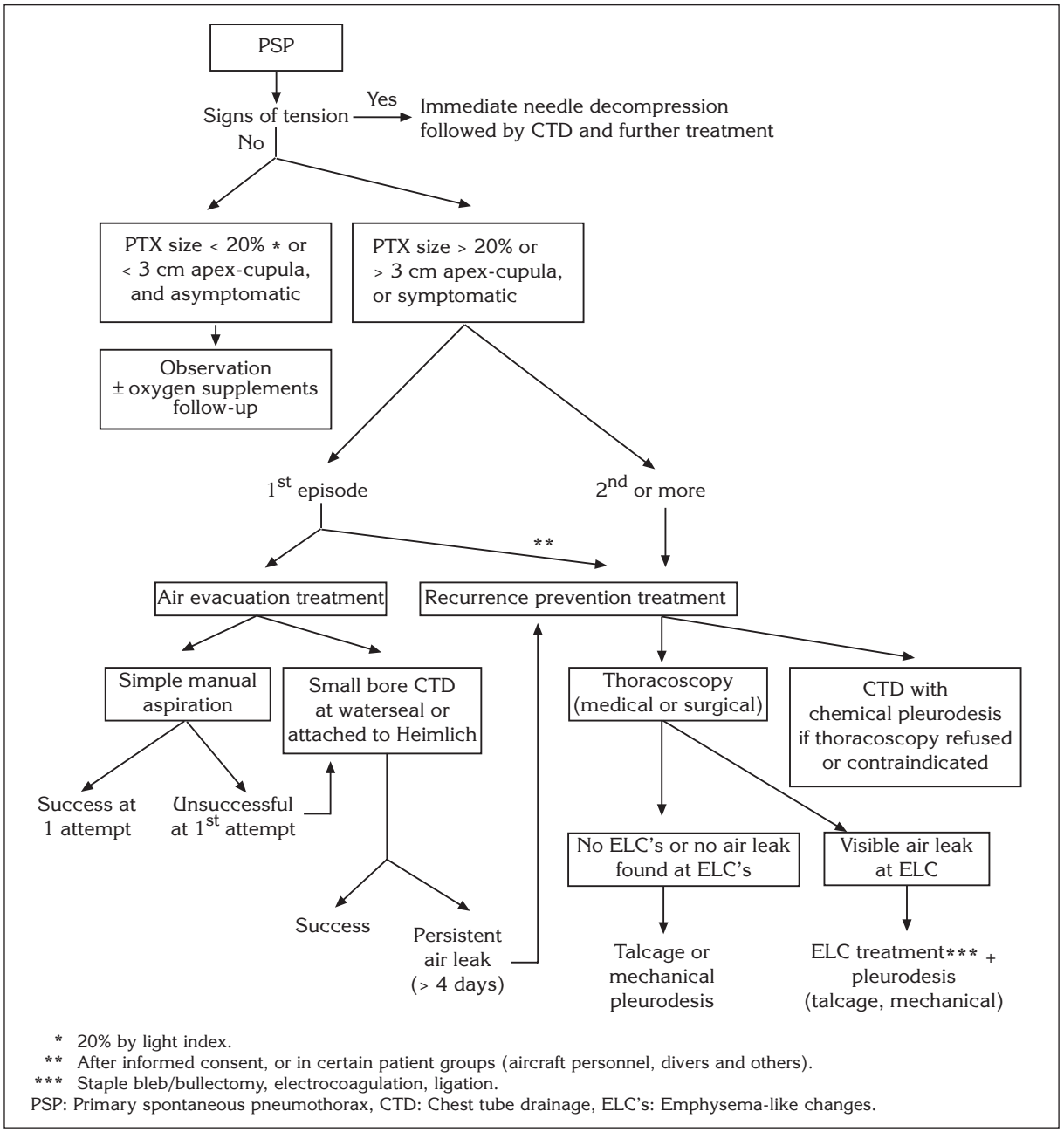

Figure 1. An algorithmic approach to the treatment of primary spontaneous pneumothorax (20).

Intercostal Catheter Drainage

Intercostal catheters usually between 10-40 F in size, it can be inserted by axillary, postero-apical and anterior approach, in recent years for practical and cosmetic reasons an axillary approach is the most favoured, success rates of 66-97% have been reported (1,27). It has some disadvantages such as chest and abdominal visceral trauma, the rate of aberrant placement is 4-9%, empyema has been estimated at 1%, another complications are bronchopleural fistulae, arteriovenous fistulae, perforation of the internal mammary artery, focal lung infections, reexpansion pulmonary edema and lung infarction (1,28).

Small bore pleural catheters (8-16 F) were as effective as large intecostal catheter resorption of PSP. A variant of small bore catheter is a pigtail catheter. Evacuation rate, total cost and mean hospital stay were similar compair with intercostal catheter drainage (1,29).

Reexpansion pulmonary edema can develops after drainage of the pneumothorax, it is a rare but potentially lethal complication. The clinical picture is dramatic and the mortality is not well defined (30). It most commonly occurs in patients with a large pneumothorax of longer duration. The pathophysiology is complex and still not completely understood.

SURGICAL MANAGEMENTS

Persistent air leak, recurrences, large bulla, spontaneous haemopneumothorax, incomplete expansion of the lung despite chest drainage and suction, tension pneumothorax, bilateral pneumothorax, in a high risk occupation (pilot and scuba diver) are include surgical indications of SP (24).

Surgical Management by Thoracotomy

Although standard posterolateral thoracotomy is still used to treat SP, the smaller incisions are preference in recent years like axillary, mini, anterior, muscle-sparing lateral and posterior thoracotomy procedures (19,31). Surgical treatment by thoracotomy used to be the last therapeutic resort for SP that could not be treated by observation, manual aspiration, drainage or thoracoscopy. The therapeutic procedure consisted of resection of lung lesions, blebs and bullae, partial or complete pleurectomy, pleural abrasion, chemical pleurodesis and the treatment of smaller bullous lesions with electrocoagulation or laser (19,32). Recurrence rates with these procedures is 3-4% (19,33). Complication rates is 0-16% (19,34). Common complications include persistent air leaks, usually defined as leaks persisting for > 5 days and occur in 5-7% of patients, wound infection 1.4-6.7%, pneumonia 2.4-8%, fever 1.9-10%, re-operation due to bleeding 1-2% and shoulder arthritis 1.9% (19,35,36). Other complications, such as residual pneumothorax, urinary tract infection, acute urinary retention, haematoma or neurological deficit are rare (19,36).

Surgical Management by VATS

VATS was first described in the surgical literature in 1992 (37,38). The experience so far shows that VATS is a safely procedure with low postoperative pulmonary complications (24,39). It can also be a useful approach in the paediatric population, high-risk and elderly patients (24). It can undergo under local and epidural anaesthesia with less access trauma (19,40). Stapled resection of bulla and talc poudrage can be performed safely (19). Patients undergoing VATS required less postoperative parenteral narcotics and pain medications than the open techniques (24,41). However, the incidence of chronic pain, numbness, or disaesthesia were present in 25-31% of patients and the rate of chronic pain was equal for VATS and thoracotomy (24,42). Recent myocardial infarction, severe coagulopathy, pleural symphysis, patients with severe lung disease and poor lung function are contraindications (24). The recurrence rate (2-14%) can be slightly higher than the mini-thoracotomy (0-7%), change to thoracotomy from VATS is required in 2% to 10% of patients with PSP and up to 29% of patients for SSP (24,43,44). Persistent air leak, wound infection, bleeding, intercostal neuralgia and emphysema are the more common complications. Rarely, re-expansion pulmonary oedema can occur (0.15%) (24,45). In SSP, treatment by VATS resulted in similar postoperative prolonged air leak and recurrence when compared with thoracotomy or PSP treated by VATS. However, the length of hospital stay was longer for SSP patients treated by VATS when compared with PSP (24,46). Post-operative complications are similar to those found after thoracotomy and occur in 1 to 27.4% of patients (19,47). So VATS is a new and discussed procedure for management of SP, most of the surgeons present (80%) did not believe that thoracotomy was required for an optimal operation and 87% believe it is more easily done by VATS. An elective operation 79% would prefer VATS to be used (37).

Pleurodesis with Sclerosing Agents and Pleural Abrassion

Pleurodesis is a management approach of SP for prevent the recurrences, it can done via chest tu-

be or thoracoscopy. Recurrence prevention should be proposed after a first recurrence, especially in patients with professional risks. After a first recurrence, recurrences increases progressively, 62% for a second recurrence and 83% for a third (19). Activation of the coagulation cascade of the pleura, fibrin deposition and fibroblast recruitment is cellular and molecular mechanisms of pleurodesis (19,48). The ideal pleural sclerosing agent should be effective, easily administered, safe, inexpensive and widely available. The most commonly used tetracyclines, silver nitrate and talc preparations. Experimental studies on animals have shown that, tetracyclines to be less efficient than talc preparations (19,49). Silver nitrate has shown superiority as a sclerosing agent in experimental studies, compared with tetracycline and talc slurry (50,51). Talc is the most inexpensive and efficient agent for pleurodesis and it was first described by Bethune in 1935 (19,52). The other pleurodesis procedure is an autologous blood patch. Autologous blood patching has been used successfully for the treatment of persistent air leak in patients with SP, it can be performed through large bore intercostal catheters (53). Pleural abrassion is an another pleurodesis procedure, it is performed by mechanical gauze abrasion, focal gauze abrasion by limited thoracotomy and mechanical abrasion by thoracoscopy using a pleural abrader (19).

Comparison Between Thoracotomy and VATS

The authors concluded that VATS was superior to thoracotomy for PSP, but doubts on its use in SSP (19). Different results have reported about the same issues in literatures, the duration of drainage was generally shorter in the VATS group or the same (19,54-56); the length of hospital or post-operative stay was also shorter or the same, the operating time was shorter for VATS, the same for both procedures, or longer for VATS than for thoracotomy, patients who underwent VATS required less narcotic analgesics than those who underwent thoracotomy, postoperative complication rates were lower or higher in the VATS group, or the same in both groups, the recurrence rate was about the same, but there seemed to be more patients with recurrences among those who underwent VATS, the amount of operative bleeding was smaller in the VATS group, patients in the VATS group returned to work earlier and finally, VATS was cheaper than thoracotomy (19,38,54-56). Most of the surgeons present (80%) did not believe that thoractomy was required for an optimal operation in contradiction to the extant guidelines and 87% believe it is more easily done by VATS (Table 2). If their own family member were to have an elective operation, 79% would prefer VATS to be used, however, in a non-elective situation the largest vote (44%) was to leave the choice of technique to the surgeon. The data for the years 2003, 2004 and 2005 show a change in practice compared with the data reported for 2000-2002, with the proportion of operations performed by VATS increasing from 57% to over 70% (Table 3) (37).

Table 2. Surgeon's opinions of thoracotomy and VATS (37).

	Agree strongly	Agree	Disagree	Disagree Strongly	Total votes	For%	Againts%
The optimal operation for pneumothorax is still through a thoracotomy	4	7	26	18	55	20%	80%
Surgery for pneumothorax is now more easily achieved by VATS	25	20	6	1	52	87%	13%
Using my technique of thoracotomy there is no advantage to the patient in having VATS	1	3	34	15	53	8%	92%

Table 3. Number of thoracotomies and VATS procedures performed by year (37).

	Thoracotomy		VATS		
Year	n	%	n	%	Total
2003	372	30	872	70	1244
2004	386	26	1077	74	1463
2005	460	27	1234	73	1694

* 37 no'lu kaynaktan alnmıştır.

CONCLUSION

The exact pathogenesis of PSP is unknown. The most common pathology is the rupture of an apical subpleural bleb, however SSP is associated with people who have lung disease, the most common cause is chronic obstructive pulmonary disease. SP can seen in some special part of life and clinical entity, such as; catamenial pneumothorax, during pregnancy, relationship with aluminum level and meteorological conditions.

Therapeutic options in cases of PSP is variations, it is include the conservative, intermediate and invasive procedure. There are two aims for treating pneumothorax: to evacuate air and to prevent recurrences. In recent years less invasive surgical techniques are prefering by surgeon and patients. Since the early 1990s, there has been a preference for VATS over thoracotomy, although the number of recurrences after VATS is generally a little higher than after thoracotomy. VATS is the only way to diminish the present confusion regarding the best treatment for pneumothorax. In conclusion we want the report a review article about pathophysiology, cilinical evaluation and treatment options of SP in the light of literatures.

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