Progresivna sustavska skleroza - promjene u orofacijalnoj regiji

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Sažetak

Progresivna sustavska skleroza generalizirana je bolest koja se manifestira i u području orofacijalne regije. Među prvim simptomima spominju se Raynaudov fenomen, napetost kože, otežano gutanje i rendgenološki zametljiv gubitak lamine dure. Liječnik stomatolog ima važnu ulogu u dijagnostici i liječenju bolesnika.

Ključne riječi: *skleroderma, orofacijalne promjene*

Acta Stomatol Croat
1999; 237—240

PREGLEDNI RAD
Primljeno: 20. lipnja 1998.

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Uvod

Usna šupljina, čeljusti i zubi često se ponašaju kao zrcalo patoloških promjena u organizmu. Zato je poznavati patologiju usne šupljine zadatak svakoga liječnika a osobito doktora stomatologije.

Sustavska skleroza ili progresivna sustavska skleroza (PSS) autoimuna je bolest karakterizirana difuznim bujanjem kolagenog vezivnoga tkiva i vaskularnim opstruktivnim promjenama koje u različitom intenzitetu zahvaćaju kožu i unutarnje organe, u prvome redu jednjak, pluća, bubrege i srce. Primarna i najčešća manifestacija bolesti počinje Raynaudovim fenomenom. Nakon toga nastaje progresivno otvrdnuće kosti koje u kasnijim stadijima može prijeći u atrofiju (1,2).

Progresivna sustavska skleroza (PSS) pokazuje simptome, kliničke znakove i rendgenološki zamjetljive promjene u orofacijalnoj regiji. U svjetskoj literaturi radile su se razne studije o promjenama orofacijalnoj regiji kod oboljelih od PSS. Klinički i rendgenski su ispitivani gubitak lamine dure, klimavost zuba, proširenje parodontalne pukotine, pojava trizmusa itd (3).

Epidemiloški PSS znatno je češća u žena. Neki autori spominju omjer oboljevanja između žena i muškaraca 4:1 (4). Procijenjeno je da prosječno u godini ima oko 2-7 novih bolesnika na milijun stanovnika (5). Bolest se najčešće javlja između trećeg i petog desetljeća života. Klinički oblik lokalizirane kožne sklerodermije (*morphea*) može se manifestirati i u drugome desetljeću života.

Bolest je nepoznate etiologije. Patogenetski se pokušava objasniti raznim teorijama o patološkim zbivanjima.

Prema vaskularnoj teoriji primarni proces događa se u malim krvnim žilama proliferacijom intime kapilara, arteriola i malih arterija, a s vremenom i većih krvnih žila. Zbog pukotine između endotelnih stanica nastaje ektravazacija plazme. Endotel podliježe destrukciji, otpuštaju se stanične organele, vrlo brzo nastaje opstrukcija lumena kapilara i bazalne lamine. Čini se da je to posljedica hipoksije. Takva proliferacija endotela kao trajnu posljedicu ima fibrozu i teleangiektazije.

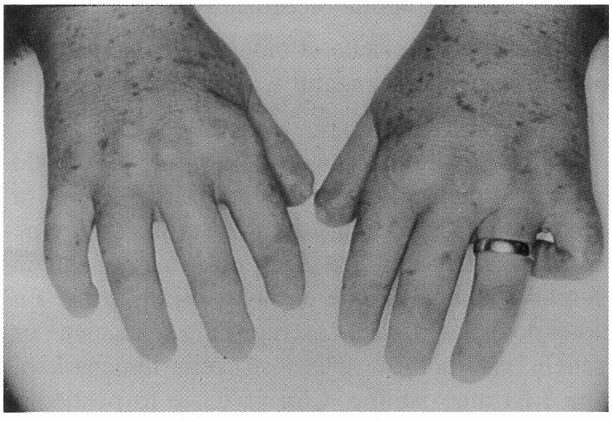
Teorija o poremećenom metabolizmu kolagena govori o primarnom procesu gubitka elasticiteta ko-

že, pa ona postaje rigidna. Pretpostavlja se da se radi o poremećenom metabolizmu kolagena, odnosno hiperaktivnoj sintezi kolagena.

Imunološka se teorija temelji na zapažanju perivaskularnih staničnih infiltrativnih promjena, dokazivanju cirkulirajućih antitijela protiv različitog tipa kolagena koji je karakterističan za stijenku krvnih žila (6). U nekih se bolesnika može naći povišen titar imunokompleksa. Prema tome, dosta elemenata govori u prilog imunološkoj teoriji.

Teorija o poremećenom autonomnom živčanom sustavu govori o povezanosti poremećaja funkcije jednjaka s Raynaudovim sindromom koji je uz sklerozu kože najvažnija klinička manifestacija PSS.

Raynaudov fenomen može mjesecima prethoditi razvoju bolesti. Glavni provocirajući čimbenik za njegov nastanak jest hladnoća okoline, a po nekim autorima i emocionalni stres (7). Važnu ulogu u nastanku Raynaudova fenomena ima poremećaj reakcije krvnih žila na hladnoću koja uzrokuje snažnu vazokonstrikciju krvnih žila subpapilarnog pleksusa. Ta vazokonstrikcija traje i nakon prestanka hladnoće, što uzrokuje blijede, hladne i ishemične prste na nogama i rukama uz bolnost. Promjene mogu biti izolirane na srednjim i distalnim falangama prstiju u obliku akrosklerotičnih promjena, pa nokti izgledaju kao kandže. Promjene na koži su dominantne, a očituju se zadebljanjem, oteklinama i crvenkastom diskoloracijom (Slika 1). Ubrzo nastaju smetnje kretanja, pa prsti postaju nepokretni. Mjestimice se javljaju teleangiektazije, tipične ulceracije, hiperpigmentacija, kožna kalcinoza, a moguća je i gangrena (8).



Slika 1. *Promjene na rukama u bolesnice koja boluje 10 godina od progresivne sustavske skleroze*

Figure 1. *Changes on hands in patient with 10 year history of progressive systemic sclerosis*

Osim na prstima, zadebljanje, tvrdoća i napetost kože mogu se širiti po čitavu tijelu. Promjene na jednjaku stvaraju disfagične smetnje u obliku otežanoga gutanja i otežanoga prolaza hrane, te osjećaja pritiska u prsnom košu. Mogu nastati i tegobe u obliku pečenja, žgaravice i boli zbog regurgitacije želučanog sadržaja i razvoja ezofagitisa. Zbog promjena na želucu i crijevima probava je poremećena slabijim prolazom hrane, nadutošću, bolovima u trbuhu, opstipacijom pa i slikom koja podsjeća na ileus (2).

Plućne promjene očituju se osjećajem nedostatka zraka, pojavom dispneje za vrijeme fizičkog napora, ali i u mirovanju. Katkada postoji suh, neproduktivan kašalj, koji je posljedica fibroze. Zbog promjena na bazalnoj alveolarnoj membrani, smanjena je difuzija plinova i kretanje prsnoga koša.

Oštećenje srčanog mišića manifestira se dekompenzacijom, aritmijama, ranim zamaranjem od kardiomiopatije zbog oštećenja srčanih mišićnih niti i fibroze (1).

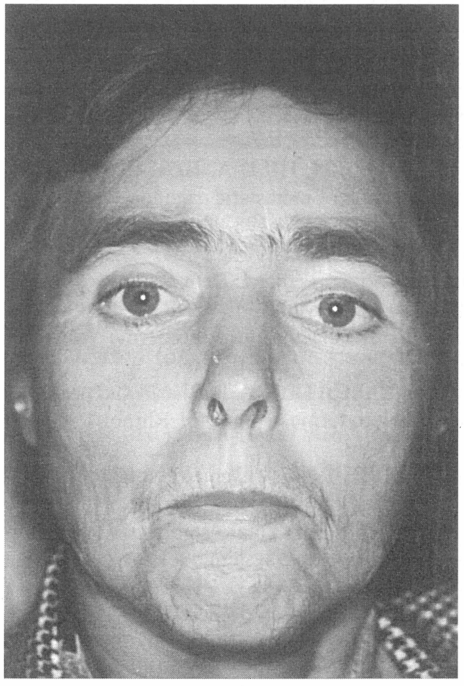
Bubrežne promjene jedan su od najčešćih uzroka smrti u bolesnika sa PSS. Manifestiraju se pojavom bjelančevina u mokraći, porastom dušićnih supstancija i naglom arterijskom hipertenzijom. U uznapredovalu stanju nalazi se slika kronične renalne insuficijencije s azotemijom, metaboličkom acidizom, poremećajem elektrolita i anemijom.

Osim opisanog oblika, PSS se može javiti u obliku CREST sindroma. U tom obliku govorimo o kalcinozi kože, Raynaudovu fenomenu, disfunkciji jednjaka, sklerodaktiliji i teleangiektaziji.

Promjene na licu i čeljusti u bolesnika sa sklerodermijom

Dugogodišnji bolesnici od sklerodermije često imaju karakterističan izgled. Postoje impresivne promjene na licu. Lice je poput maske, koža je napeta, bez nabora, a mimika je ograničena ili je nema. Lice je glatko sjajno s malim ušiljenim nosom. Sluznica usana se sužava, elasticitet je smanjen, a poslije se otvor usta smanji pa nastaje mikrostomija. Na taj je način funkcija usana smanjena. Karakterističan je simptom pojava teleangiektazija na licu ili u težim slučajevima nastanak pe-

riorbitalnog hematoma (9,10). U 40-50% slučajeva promjene zahvaćaju jezik. On se smanji, postaje gladak, a frenulum se skrati (Slika 2).



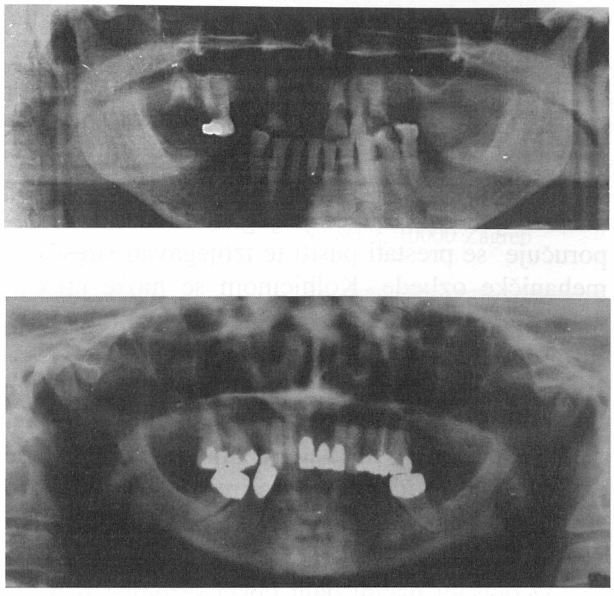
Slika 2. *Izgled bolenice koja boluje od PSS 8 godina*
Figure 2. *Patient with 8 year history of PSS*

Ograničeno otvaranje usta često je udruženo ne samo sa kontrakturom tijekom otvaranja usta nego i s pseudoankilozom temporomandibularnog zgloba (11).

Atrofija muskulature jezika i mekog nepca, resorpcija alveolarne kosti, resorpcije angulusa mandibule, destrukcija kondila, deficitaran zigomatični luk, atrofije masetera i pterigoidnih mišića, skraćivanje i napetost u području frenuluma jezika, pojava kalcifikacija u mekom tkivu usne šupljine ili pojava otvorena zagriza spadaju u simptome koji prate PSS (12-20) (Slika 3).

Weber (21) opisuje obostranu frakturu čeljusti u bolesnika sa sklerodermijom koja je posljedica pada, ali je povezana s pojačanom resorpcijom čeljusti zbog osnovne bolesti.

Zbog navedenih promjena u bolesnika s PSS održavati higijenu usne šupljine je otežano. Ograničena pomoć je stalna i dobra oralna higijena. Kod zahvata u stomatološkoj ordinaciji dolaze u obzir



Slika 3. *Ortopantomogram u dvije bolesnice koje boluju od PSS 10 godina*
Figure 3. *Panoramic radigraph of two female patients with 10 year history of PSS*

svi konzervativni zahvati koji se mogu učiniti te eventualne ekstrakcije, ako su jedini izbor u liječenju. Opasnost kod zahvata je laceracija komisure usana. Literatura spominje i komisurotomiju kao jedan od oblika liječenja sekundarne mikrostomije te način za eventualno protetsko zbrinjavanje (22). Periodičnim kliničkim pregledom i eventualnim rtg snimkama moguće je u određenoj mjeri pomoći pacijentima kako bi se izbjegle ili eventualno umanjile posljedice koje nastaju progresijom bolesti.

Dijagnoza se u većini slučajeva može postaviti na osnovi kliničke slike i anamnestičkih podataka. Među rane simptome treba ubrojiti hladne prste, napetu kožu na distalnim falangama, smetnje prstiju prigodom kretanja, Raynaudov fenomen, napetost kože lica, ekstremiteta i trupa, te smetnje pri gutanju (23,24). Od laboratorijskih pretraga histološki pregled kože pokazuje homogenizaciju kolagena. Rendgenskom snimkom pluća nađu se intersticijske promjene, a spirometrijom smetnje restriktivnog tipa. Rendgenski pregled jednjaka pokazuje proširen, mlohav sa slabom peristaltikom. EKG može pokazivati smetnje provođenja. U krvi se mogu naći antinuklearni čimbenik, krioglobulin, reumatoidni čimbenik, hipergamaglobulinemija i povišen titar imunokompleksa.

Do danas još nije poznato etiološko liječenje, niti se uspjelo bitno utjecati na tijek bolesti. U terapiji se spominju glukokortiokoidi, preparati penicillina, citostatici, indometacid (25 Dominantne promjene na koži mogu se liječiti raznim kremama, mastima za kožu, kupkama te aktivnim fizikalnim vježbama. Treba izbjegavati hladnoću. Preporučuje se prestati pušiti te izbjegavati stresove i mehaničke ozljede. Kolhicinom se može utjecati da se smanji kolagen u koži te eventualno poboljša motorika prstiju. Ako postoji renalna hipertenzija, upotrebljavaju se antihipertenzivi, a nifedipin i drugi blokatori kalcija mogu pomoći ublažiti Raynaudov sindrom. Eventualni ezofagitis i druge vrste probavnih smetnji liječe se primjenom ranitidina i cimetidina te konzumiranjem hrane u malim i kašastim obrocima.

Svi opisani načini daju dosta skromne rezultate. Bolest je kroničnoga karaktera, uglavnom progresivnog polaganog razvoja, premda je moguć i brz napredak bolesti. S vremenom tegobe postaju sve jače, a pokretljivost slabija.

Uzroci smrtnoga ishoda jesu bronhopneumonija, perforacije u gastrointestinalom traktu, srčana, plućna ili renalna insuficijencija često udružena s malignom hipertenzijom (1).

Rasprava i zaključak

U dijagnostici i liječenju progresivne sustavske skleroze nisu iskorištene sve mogućnosti. Bolesnici oboljeli o skleroderme postaju sve veći medicinsko-društveni problem te je uloga liječnika stomatologa u prevenciji, dijagnostici i terapiji nezaobilazna. Zato bi bilo poželjno ustrojiti multidisciplinirani pristup u dijagnostici i terapiji bolesnika od progresivne sustavske skleroze.

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Progressive Systemic Sclerosis - Changes in the Orofacial Region

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Summary

Progressive systemic sclerosis is a generalised disease which also manifests in the orofacial region. Among the first symptoms are Raynaud's phenomena, tense skin, difficulty swallowing and radiologically visible loss of lamina dura. The dentist has an important role in the diagnostics and treatment of this disease.

Key words: *scleroderma, orofacial changes*

Acta Stomatol Croat
1999; 241—243

REVIEW
Received: June 20, 1998

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Introduction

The oral cavity, jaws and teeth frequently reflect pathological changes in the organism. For this reason knowledge of the pathology of the oral cavity is the primary task of every physician, particularly the dentist.

Systemic sclerosis or progressive systemic sclerosis (PSS) is an autoimmune disease, characterised by diffuse swelling of the collagen of connective tissue and vascular obstructive changes, which to various degrees of intensity involve the skin and internal organs, primarily the oesophagus, lungs, kidneys and heart. The principal and most frequent manifestation of the disease begins with Raynaud's phenomena, followed by progressive hardening of bones and in the later stages atrophy may occur (1,2).

Progressive systemic sclerosis (PSS) presents symptoms, clinical signs and visible radiological changes in the orofacial region. According to world literature various studies have been performed on changes in the orofacial region in patients with PSS. Loss of lamina dura, loose teeth, widening of the parodontal gap, occurrence of trismus, etc. have been studied both clinically and radiologically (3).

Epidemiologically, PSS is significantly more frequent in women, and some authors suggest a ratio between women and men of 4:1 (4). It is estimated that each year 2-7 new patients occur per one million inhabitants (5). Manifestation of the disease most often occurs between the third and fifth decade of life. The clinical form of localised skin scleroderma (morphea) can manifest in the second decade of life.

The disease is of unknown aetiology. Attempts have been made to explain it pathogenetically by various theories of pathological occurrences.

According to the vascular theory the primary process takes place in the small blood vessels, with proliferation of the intima of capillaries, arteriole and small arteries, and later larger blood vessels. Because of gaps between the endothelial cells plasmic extravasation occurs. The endothelium is subjected to destruction, cellular (organele) are rele-

ased and rapid obstruction of the capillary lumena and basal lamina occurs. Which appears to be a consequence of hypoxia. Such proliferation of the endothelia results in permanent fibrosis and telangiectasia.

The theory of disturbed metabolism of collagen suggests that the primary process is loss of elasticity of the skin, which becomes tense. It is hypothesised that this is caused by disturbed metabolism of collagen, i.e. hyperactive synthesis of collagen.

The immunologic theory is based on observation of perivascular cellular infiltrative changes, detection of circulating antibodies against a different type of collagen, which is characteristic for the blood vessel wall (6). In some patients raised titar for immunocomplex can be found. Thus, several elements favour the immunologic theory.

The theory of disturbed autonomic nervous system assumes a connection between oesophagus function and Raynaud's syndrome, which, apart from sclerosis of the skin, is the most significant clinical manifestation of PSS.

Raynaud's phenomena may precede development of the disease for months. The main provoking factor for its occurrence is a cold environment, and according to some authors emotional stress (7). An important role in the occurrence of Raynaud's phenomena is disturbed reaction of the blood vessels to cold, causing powerful vasoconstriction of the subpapillary plexus blood vessels. This vasoconstriction continues even after cessation of cold, resulting in pale, cold, ischemic digits (fingers and toes) and pain. Changes may be isolated to the middle and distal phalanges of the digits, in the form of acrosclerotic changes, with nails which resemble claws. Skin changes are dominant with thickening, swelling and reddish discoloration (Figure 1). Disturbed movement quickly develops and the digits become immovable. In places there is telangiectasia, typical ulceration, hyperpigmentation, calcinosis cutis, and possible gangrene (8).

Apart from the digits, the skin may become hard and tense over the whole body. Changes on the oesophagus lead to dysphagic disturbances, such as difficulty swallowing and difficult passage of food, and a sensation of pressure in the chest. Other disturbances may occur in the form of a burning sensation, heartburn and pain, due to regurgitation of the stomach contents and development of oesophagitis. Because of the changes in the stomach and intestines digestion is disturbed by the poor passage of food, distention, abdominal pain, constipation and a condition which resembles illeus (2).

Pulmonary changes manifest with a sensation of lack of air, dyspnea during physical exercise and rest. Occasionally there is a dry, non-productive cough, as a consequence of fibrosis. Because of changes on the basal alveolar membrane, gas diffusion and chest movement are reduced.

Damage to the heart muscle manifests with decompensation, arrhythmia, premature fatigue as a result of cardiomiopathy, due to damaged heart muscle fibres and fibrosis (1).

Kidney changes are one of the most frequent causes of death in patients with PSS. They manifest with the occurrence of proteinuria, increased nitrogenous substances and sudden arterial hypertension. The advanced stage presents with chronic renal insufficiency and azotemia, metabolic acidosis, disturbed electrolytes and anaemia.

Apart from the aforementioned form of PSS it can also occur in the form of CREST syndrome, in which there is calcinosis cutis, Raynaud's phenomena, oesophagus dysfunction, sclerodactyly and teleangiectasia.

Changes on the face and jaws in patients with scleroderma

Patients with longterm scleroderma often have a characteristic appearance, and impressive changes are noticeable on the face. The face may have the appearance of a mask when the skin is tense without wrinkles, and mimic is limited or absent. The face is smooth, shiny and the nose small and pointed. The mucous membrane of the lips is contracted, elasticity is reduced and the opening of the mouth becomes progressively smaller until microstomia occurs and lip function is reduced. A characteristic symptom is the occurrence of facial telangiectasia, or in more serious cases the occurrence of periorbital haematoma (9,10). In 40-50% of cases changes include the tongue, which beco-

mes smaller, smooth and the frenulum shorter (Figure 2).

Restricted opening of the mouth is not only accompanied by contracture during mouth opening, but also very often by pseudoankylosis of the temporomandibular joint (11).

Atrophy of the tongue and soft pallet musculature, resorption of alveolar bones, resorption of the mandibular angulus, destruction of the condyle, deficient zygomatic arch, atrophy of the masseter and pterygoidic muscles, shortening and tension in the area of the tongue frenulum, calcification in the soft tissue of the oral cavity or the occurrence of open bite, are included in the symptomatology which accompanies PSS (12-20) (Figure 3).

Weber (21) described bilateral fracture of the mandibula in a male patient with scleroderma, resulting from a fall, connected with increased resorption of the mandibula as the outcome of the basic disease.

Because of the aforementioned changes in patients with PSS maintenance of oral cavity hygiene is hindered. Constant, adequate oral hygiene is of limited help. With regard to operations in the dental surgery, all possible conservative operations can be considered, including eventual extractions if they are the only choice of treatment. With regard to operations, there is a danger of laceration of the commissura of the lips. It has been reported in literature that commissurotomy is one form of treatment for secondary microstomy, and a means for eventual prosthetic care (22). A periodic clinical examination and eventual X-ray may, to a certain extent, help patients to avoid or eventually diminish the consequences of progression of the disease.

In the majority of cases diagnosis can be made on the basis of the clinical status and history data. Among the early symptoms the following should be considered: cold fingers with tense skin on the distal phalanges, impaired digital movement, Raynaud's phenomena, tense facial skin, extremities and trunk, and difficulty swallowing (23,24). With regard to laboratory examinations histologic examination of the skin shows homogenisation of collagen. A chest X-ray shows interstitial changes, and spirometry shows restrictive type disturbance. An X-ray of the oesophagus reveals an extended, flaccid oesophagus with poor peristaltic. An ECG may show disturbed conduction. Blood analysis may show antinuclear factor, crioglobulin, rheumatoid factor, hypergammaglobulinemia and raised titar immunocomplex.

Etiologic treatment is still not known and neither has it been possible to essentially effect the course of the disease. With regard to therapy mention is made of glucocorticoides, penicillin preparations, cytostatics, indometacid (25). Dominant changes on the skin can be treated with various creams, ointments, washes and physical exercise. Cold should be avoided. It is recommended that patients should stop smoking, avoid stress and mechanical injury. Kolhicin can have an effect on a reduction of collagen in the skin and eventual improvement in digital motorics. In the case of renal hypertension antihypertensives are used, while nifedipin and other calcium blocking agents can help to alleviate Raynaud's syndrome. Oesophagitis and other digestive disorders are treated by alternating ranitidin and cimetidin, and consumption of food in small, mashed portions.

However, all the above methods produce relatively poor results. The disease is of a chronic nature, usually with a slow, progressive course, although rapid advancement of the disease can occur. In time the disorders become more marked and movement poorer.

The causes of lethal outcome are bronchopneumonia, perforation of the gastrointestinal tract, cardiac, pulmonary or renal insufficiency, which is frequently accompanied by malignant hypertension (1).

Discussion and conclusion

All possibilities for diagnostics and treatment of progressive systemic sclerosis have not been utilised. Patients with scleroderma are becoming a significant medical-social problem and the role of the dentist in the prevention, diagnostics and therapy is indisputable. Thus, a multidisciplinary approach in the diagnostics and therapy of patients with progressive systemic sclerosis should be determined.